TEXTBOOK OF BRITISH SURGERY

Edited by

Sir HENRY SOUTTAR, C.B.E., D.M.(Oxon.), F.R.C.S.

[illegible]

and

J. C. GOLIGHER, Ch.M.(Edin.), F.R.C.S.(Eng. and Edin.)

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VOLUME TWO: THE CENTRAL NERVOUS SYSTEM—THE EYE—EAR, NOSE AND THROAT—MOUTH SALIVARY GLANDS AND JAWS NECK THE BREAST—THE ŒSOPHAGUS—THE HEART AND LUNGS



1957

WILLIAM HEINEMANN · MEDICAL BOOKS · LTD

LONDON

First Published 1957

Printed in Great Britain by the Pitman Press, Bath

EDITOR'S PREFACE

THE advances of Surgery in the last twenty years have been so great that no one individual can master all the fields which they have opened. On the other hand it is important that candidates for the higher examinations should be familiar with the whole subject, for only thus will they be able to select for their future career the branch for which they are best adapted. This new *Textbook of British Surgery* aims to meet their requirements.

It has been compiled by more than forty authors, each an acknowledged master in his own particular branch, and the aim has been to give a clear and succinct but complete account of the present position in each field. So rapid has progress in surgery been that several of these articles have been completely rewritten while the book was being compiled. At the moment they present an accurate view of surgical practice today on its highest plane. We hope that they will be of material use to the student in acquiring the knowledge which is necessary for his work, and that later on they may inspire him to add to that knowledge and by his own labours to develop still further the great subject to which he is devoting his life.

H. S.
J. C. G.

London, 1957

GENERAL PREFACE

This volume of the *Textbook of British Surgery* covers The Central Nervous System —The Eye—Ear, Nose and Throat—Mouth Salivary Glands and Jaws—Neck—The Breast—The Œsophagus—The Heart and Lungs, discussing fully diagnosis, pathology, prognosis and treatment of the areas concerned.

Sir Henry Souttar, as General Editor, has now had the assistance of Professor J. C. Goligher of Leeds, and between them they have collected in Volume II a team of recognized authorities on the subjects treated. In a work of multiple authorship such as this there are inevitably occasional differences of opinion and some overlapping. We have deliberately accepted this as adding to the interest of the subjects discussed. The standard aimed at is as complete an account as possible of the present position in each field of surgery as seen by the expert to whom the section has been entrusted. The names of the contributors are a sufficient guarantee that this standard has been attained. They are drawn from the leading medical schools and hospitals throughout Britain and the Commonwealth and represent the consensus of opinion in present-day British Surgery.

The volume is essentially clinical and practical, with such pathology as is necessary for diagnosis and treatment. Surgical procedures are described, and the authors discuss the advantages and disadvantages of each procedure in vogue, indicating their reasons for preferring one to another.

The volume is illustrated by original drawings, photographs, X-rays, and diagrams. The audiences it is intended to interest are general surgeons, registrars, postgraduate students and those reading for the Fellowship and other higher examinations.

LIST OF CONTRIBUTORS

JOHN ANDERSON, M.B., F.R.C.P.,
Physician, St. Thomas's Hospital, London.

N. R. BARRETT, M Chir., M.B., F.R.C.S. (Eng.),
Surgeon, St. Thomas's and The Brompton Hospitals, London.

A. G. CROSS, M.D., F.R.C.S (Eng.),
Consulting Ophthalmic Surgeon, St. Mary's Hospital, London and Moorfields Eye Hospital, London.

CHARLES DONALD, O.B.E., F.R.C.S. (Eng.),
Late Surgeon, London Hospital.

DAVID EVANS, M.B., F.R.C.S. (Eng),
Consultant Surgeon, S E. Kent Group of Hospitals, formerly Senior Surgical Registrar, London Hospital.

M. R. EWING, M.B., F.R.C.S. (Edin. and Eng.),
Professor of Surgery, Melbourne University, Australia.

R. H. FRANKLIN, M.B., F.R.C.S. (Eng.),
Consulting Surgeon, Kingston and Hammersmith Hospitals, Senior Lecturer in Surgery, Postgraduate Medical School, London.

R. S. HANDLEY, O.B E., M.B., F.R.C.S (Eng.),
Surgeon, Middlesex Hospital, London.

C A. KEOGH, M.B., F.R.C.S. (Eng.),
Surgeon, Ear, Nose and Throat Department, London Hospital.

CHARLES LANGMAID, M.B. (Lond.), M.B., B.Sc. (Wales), F.R.C.S. (Eng.),
Consulting Surgeon, Neurosurgery, United Hospitals, Cardiff.

J. E. PIERCY, F.R.C.S. (Edin and Eng.),
Surgeon Superintendent, Thyroid Unit and Department of Endocrinology, New End Hospital, Hampstead.

LAMBERT C. ROGERS, V.R.D., M.D., M Sc., F.R.C.S (Edin. and Eng), F.R.A.C.S., F.A.C.S.,
Professor of Surgery, University of Wales, Director of Surgical Unit, Cardiff Royal Infirmary; Surgeon, United Cardiff Hospitals, Consultant in Neurosurgery to the Royal Navy

T. HOLMES SELLORS, D.M., M.Ch (Oxon.), F.R.C.S. (Eng.),
Surgeon, National Heart Hospital and London Chest Hospital, Thoracic Surgeon, Middlesex Hospital.

DILLWYN M. E. THOMAS, F.R.C.S. (Eng.),
Consulting Thoracic Surgeon, United Cardiff Hospitals and Welsh Regional Hospital Board.

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CHAPTER I

THE CENTRAL NERVOUS SYSTEM

INTRODUCTION

LAMBERT ROGERS and CHARLES LANGMAID

THE SCALP AND THE SKULL

Then mark the cloven sphere that holds
All thought in its mysterious folds,
That feels sensation's faintest thrill,
And flashes forth the sovereign will,
Think on the stormy world that dwells
Locked in its dim and clustering cells!
The lightning gleam of power it sheds
Along its slender glossy threads!

OLIVER WENDELL HOLMES

THIS account of the surgery of the central nervous system is largely a record of our experience of its practice but we have not hesitated to draw on the literature to amplify this wherever necessary. Details of operative technique are not given and the practical rather than the more academic aspects of injuries and diseases of the nervous system are stressed.

The historical evolution of modern neurosurgical practice is not here considered but we should never forget the great advances which have made it possible and the high priests who introduced them, such as Horsley, Macewen, Cushing, Dandy and Moniz. The two last by their discovery respectively of ventriculography and angiography made the localization and recognition of intracranial tumours an exact science from being merely speculation, while Forestier and Sicard did the same for space-occupying lesions of the spinal canal. Finally we should never forget Harvey Cushing's oft-repeated precept,

"Don't let neurological surgery get too far away from general surgery."

Sound principles are the secret of success in surgery whatever the region of the body in which it is performed and as has been rightly said, "no excellence of tactics will correct a fault in strategy."

Between the pericranium of the skull bones and the epicranius or occipito-frontalis muscle and its aponeurosis, the galea aponeurotica, lies the sub-aponeurotic potential space in the plane of which the scalp moves over the skull Connected by veins to the intracranial venous sinuses this space is a potentially dangerous area inasmuch as infection may spread from it within the skull and give rise to cavernous or other sinus thrombosis and complications. Scalp wounds extending into the space therefore must never be regarded as trivial and although the relatively rich blood supply of the scalp is conducive to their rapid healing and an infrequency of infection, they should nevertheless be given early and careful surgical attention.

Because developments in radiography, such as ventriculography and angiography,

have given us exact methods of defining relationships and since bone flaps are reflected when it is desired to examine extensive areas of the brain, the necessity to mark out various base lines and use particular points in order to establish sites of exposure no longer exists. It is well to remember, however, the approximate position of the central (Rolandic) fissure of the brain. This lies approximately beneath the upper 3 or 4 in. of a line drawn downwards and forwards towards the tubercle of the zygoma from a point half an inch behind the mid-point of the *nasion-inion line*. Another point which may be of value, especially in emergency, is the position of the anterior branch of the middle meningeal artery. This lies under a point 1½ in. above the zygoma and 1½ in. behind the external angular process of the frontal bone.

THE BLOOD SUPPLY OF THE BRAIN AND CORD

The brain of man receives blood from the two internal carotid arteries* and the two vertebrals which unite to form the basilar. About three-quarters of the brain is supplied by the carotids, the remainder by the vertebrals, the fore-brain and mid-brain being supplied by the carotids, the hind-brain, the medulla and spinal cord, by the phylogenetically older vertebral system.

The presence of S-shaped flexures in the internal carotids and vertebrals which are peculiar to these vessels allows movements of the head to take place without disturbance of the blood flow in their channels.

The Internal Carotid

The point of origin of the internal carotid artery is of some practical importance. It is usually stated as at the level of the upper border of the thyroid cartilage but it is in reality very variable. It is very rarely below this level but as angiography has recently confirmed is frequently above it and may lie beneath the angle of the mandible and as high as the 2nd cervical vertebra or even the atlas. These variations occur more often in women than in men.

In the neck the internal carotid is devoid of branches. As it enters the skull it is peculiar inasmuch as it fits the carotid canal within the petrosal bone rather closely and nowhere else in the body does a large artery have a comparable intra-osseous course. In the canal it is surrounded by a plexus of small veins, is accompanied by sympathetic fibres from the superior cervical ganglion and gives off its tympanic branch. It is also peculiar in having an intracranial course within a venous sinus. Within the cavernous sinus the artery itself and numerous interlacing filaments of fibrous tissue which support it, are covered by the endothelium of the sinus. As it passes through the sinus the artery makes an *S*-shaped bend, the carotid siphon.

While in the cavernous sinus the internal carotid gives off small branches to the hypophysis and the Gasserian ganglion: the ophthalmic artery has its origin just as the main artery leaves the sinus. At this point the calibre of the carotid is only about one-quarter of the size of the vessel in the neck.

At the base of the brain, having perforated the roof of the cavernous sinus and given off its ophthalmic branch, it gives off the Posterior Communicating and Anterior

* "Carotids or soporales, the sleepy arteries, because they being obstructed, or in any way stopt, we presently fall asleep."

Ambroise Paré.

Choroidal arteries and divides into its terminal branches, the Anterior and Middle Cerebrals.

The Anterior Cerebral

The anterior cerebral gives off three sets of branches from (*a*) its convexity, (*b*) its concavity, (*c*) basal. Of the latter an important one is the artery of HEUBNER which passes through the anterior perforated spot to the caudate nucleus, anterior part of the putamen and the anterior limb of the internal capsule.

Variations. The anterior cerebral artery may be a simple azygos vessel or it may be duplicated on one hemisphere. *The anterior communicating artery* which is very constant in man is absent in some animals.

The Middle Cerebral or Sylvian Artery

This is the largest branch and appears to be the direct continuation of the internal carotid. It lies in the Sylvian fissure and its main branches as seen in an angiogram mark the upper border of the temporal lobe. Emboli in the internal carotid tend to pass into the middle cerebral. The direct course of an embolus in the aorta is into the left common carotid rather than back through the innominate into the right common carotid and so it passes to the left middle cerebral, producing right hemiplegia and aphasia in the right-handed. The main branches of the middle cerebral artery are (*a*) ascending, (*b*) descending, (*c*) perforating. Of the latter the lenticulo-striate artery of DURET (the artery of cerebral hæmorrhage of CHARCOT) is important.

The anterior choroidal artery passes backwards on the medial surface of the temporal lobe, entering the inferior horn of the lateral ventricle through the choroidal fissure to supply the choroid plexus. Its course is apparent in about 30 per cent of carotid angiograms (Mounier-Kuhn *et al.*, 1955).

The Posterior Cerebral

The two posterior cerebral arteries arising as the termination of the basilar artery in the ponto-mesencephalic sulcus, course round the side of the brain stem and continue into the depths of Bichat's fissure. These vessels arise originally as branches of the internal carotid and fuse with the termination of the basilar. Each posterior cerebral terminates by dividing into (*a*) an inferior temporal branch to the cuneus and the convex surface of the hemisphere, (*b*) an occipital or calcarine artery to the calcarine fissure, cuneus and precuneus. Near the origin of the posterior cerebral, lying between this artery above and the superior cerebellar artery below, the third nerve passes forwards from its origin. The posterior cerebrals and their branches form a complete arterial circle around the brain stem and a vascular plexus (the quadrigeminal plexus) is formed with branches of the superior cerebellar arteries.

The posterior choroidal artery from the posterior cerebral enters the choroidal fissure to supply the choroid plexuses of the third and lateral ventricles. Angiography may show displacement of the vessel in intraventricular meningiomas (Wall, 1954).

The Vertebral

In over 90 per cent of subjects the vertebral arteries are unequal in size, neither side constantly preponderating. The development of the artery as a preneural, post-costal longitudinal anastomosis may explain this variability.

The anterior spinal artery and the posterior inferior cerebellar artery are both branches of the vertebral. The posterior spinal artery usually arises from the posterior inferior cerebellar (75 per cent of cases) but otherwise arises from the vertebral.

The Circle of Willis

The circle (more often called on the Continent, the heptagon or polygon) of Willis (Fig. 1) is formed by the two anterior cerebrals and their connexions, the anterior communicating artery, the two posterior cerebral and the posterior communicating



(From "Brain," Macmillan & Co Ltd)
FIG 1 Dissection of circle of Willis from a man aged 48 The choroid plexuses from the lateral ventricles can be seen

arteries and the internal carotids. Although the arterial ring is frequently asymmetrical, it is very rarely defective and its function appears to be that of an anastomosis, the connecting vessels of which are capable of opening up if required, and not that of an "equalizer" of the blood supply to the brain as has been thought by some (Rogers, 1947)

The Inner Circle

Small arterial twigs emerge from the bifurcation of the basilar artery and meet in a

These fine tufts have been

The Blood Supply of the Cerebellum

Consequent upon the expansion of the lateral cerebellar masses typical of the cerebellum of the primates and of man, the posterior inferior cerebellar artery achieves preponderance over the superior cerebellar which is the larger artery in most mammals.

End Arteries

The cortical ramifications of the arteries to the cortex were at one time regarded as end arteries but this is not so as these vessels communicate freely with each other over the surface of the brain. The vessels which penetrate the cerebral substance, however, do not as a rule anastomose and for the most part are true end arteries.

Vaso-Motor Control

The common and internal carotids are richly beset with nerve fibres of the sympathetic system and despite denials to the contrary the cerebral vessels are under the influence of vaso-constrictor and vaso-dilator impulses. Forbes and Cobb of Boston (1938) have clearly demonstrated the presence of vaso-constrictor fibres but have shown that they are only one-tenth as effective on the vessels in the pia as are the vaso-constrictor nerves on the vessels in the skin. The intra-cerebral regulation of blood flow is chiefly by means of chemical agents, e.g. carbon dioxide.

Venous Drainage

The venous drainage of the brain is through the internal jugular veins, the right being the larger in most subjects, in whom the superior sagittal (longitudinal) sinus is continued at the torcular into the right transverse sinus and so into the right internal jugular.

The superficial cerebral veins include the superior cerebral veins joining the superior longitudinal sinus. These vary in number and as a rule course in a somewhat parallel fashion towards the superior sagittal (longitudinal) sinus. The superficial group includes the anastomosing veins of LABBÉ and TROLARD.

The deep cerebral veins draining the basal ganglia and choroid plexus include the choroid veins, the vena terminalis or thalamo-striate vein, the vein of the septum pellucidum and the internal cerebral vein.

The basal vein of ROSENTHAL begins at the anterior perforated substance and is formed by the union of the anterior cerebral veins, the deep SYLVIAN vein and the inferior striate vein. It runs backwards and upwards in the cisterna ambiens around the cerebral peduncle and with its fellow of the opposite side joins with the two internal cerebral veins to form the great vein of GALEN. These veins are well shown in the venous phase of an angiogram (phlebogram) and their various displacements may be of great help in the diagnosis of hemisphere tumours.

The great vein of Galen passes backwards and upwards and then turns slightly forwards around the splenium of the corpus callosum to join with the inferior sagittal sinus and form the straight sinus which courses through the tentorium cerebelli to the confluence of the sinuses at the torcular HEROPHILI. These latter vessels are fixed by the rigid falx and tentorium and are not likely to suffer any displacement (Lin *et al.*, 1955) (Johanson, 1954).

Small veins pass across the subdural space from the cerebral cortex to enter the superior longitudinal sinus. Others pass from the cerebellar cortex to enter the

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FIG 1 Dissection of circle of Willis from a man aged 48 The choroid plexuses from the lateral ventricles can be seen

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The Inner Circle

Small arterial twigs emerge from the bifurcation of the basilar artery and meet in a transverse anastomosis in the fossa interpeduncularis constituting a small vascular circle within the circle of Willis. Fine hair-like vessels leave the constituent vessels of the circle of Willis to supply the hypophysis and other structures within the interpeduncular space. These fine tufts have been known as Monakow's Vascular Beard.

the cord is sometimes helpful to the surgeon in enabling him to decide whether a laminectomy is above or below the site of the cord compression; the blood flow in these veins is upwards and so those below the site of compression are prominent and congested while those above appear normal. The veins of the spinal canal are large and complex in their arrangement, but for the most part form anterior and posterior plexuses related respectively to the backs of the vertebral bodies on either side of the posterior common ligament, and to the deep surfaces of the laminæ and ligamenta subflava. Wide channels connect the veins of the spinal canal with dorsal spinal venous plexuses in the vertebral grooves. Branches also traverse the intervertebral foramina to join the posterior branches of the intercostal and lumbar veins. At the base of the skull the venous plexuses enter the basilar and occipital sinuses and give off branches which emerge above the posterior arch of the atlas to form the origin of the vertebral vein.

Pressure on the abdomen causes congestion of the veins within the spinal canal which may be the cause of troublesome bleeding in certain operations, e.g. for intervertebral disk lesions. This congestion may be avoided by so arranging the patient on the operation table that the body weight is taken by the iliac crests and not by the abdomen.

THE MENINGES AND THE CEREBROSPINAL FLUID

The brain and cord are enclosed in the dura mater which is closely applied to the skull bones but is separated from the laminæ and ligamenta flava of the spinal canal by a varying amount of epidural fat which is usually bluish in colour and is occasionally the source of a lipoma which may compress the spinal cord. Within the dura is the avascular arachnoidea enclosing the cerebrospinal fluid, movements of which have been described by Harvey Cushing as the third circulation. There is a self-insulating mechanism of the nervous system to which Wilfred Trotter (1926) drew attention and which can be seen and verified by anyone who examines it. If cerebrospinal fluid is allowed to come into contact with somatic tissues such as muscle it becomes shut off from it by the formation of a "new" dura in the form of a glistening smooth membrane which forms to enclose it. This is a very curious phenomenon inasmuch as cerebrospinal fluid is principally water containing small amounts of salts and protein and how it is brought about is unknown. It is well seen after a laminectomy has been performed and the dura left unsutured. If a second operation is performed a smoothly lined sac containing cerebrospinal fluid will be found extending between the sacro-spinales at the site of the defect.

Intrameningeal Pressure

In measuring the pressure of the cerebrospinal fluid at lumbar puncture it must be remembered that this is affected by the height of the skull above the horizontal. When cisternal puncture is performed with the patient sitting in a chair with the head flexed it is often necessary to aspirate the fluid from the cisterna magna as the pressure within it in this position may be below atmospheric. When the intracranial cerebrospinal fluid pressure is raised papillœdema is apparent on ophthalmoscopic examination. The quantity of the fluid (which is incompressible) materially affects the total intracranial pressure and does so more than any other of the cranial contents. Thus a small tumour in the posterior fossa by obstructing the iter and/or congesting the choroid plexuses by distorting the straight sinus in the tentorium cerebelli and thereby obstructing the great vein of Galen, may produce a great rise in intracranial pressure, whereas a much larger

lateral sinus. These "bridging" veins are liable to rupture in head injuries and give rise to subdural hæmatomas. The cerebral "bridging" veins are particularly liable to give way if the brain is shaken antero-posteriorly such as occurs when a blow is delivered to the front or back of the skull. In laterally delivered blows the falx acts as a buffer and limits the degree of movement of the brain and the consequent liability of the veins to be torn.

The Middle Meningeal Artery and Veins

This artery is of importance to the surgeon because of its liability to rupture in cases of head injury and give rise to extradural clot compression of the brain (see p. 16). The artery having entered the skull through the foramen spinosum has a short course of from ¼–1 in. in length and lies in a groove on the inner table of the squamous temporal bone. It then divides into its two terminal branches, the anterior and larger of which runs upwards across the anterior-inferior angle of the parietal bone and ascends towards the vault of the skull a short distance behind the coronal suture; the posterior branch runs almost horizontally backwards across the squamous temporal bone and takes the line of the second temporal convolution. The trunk of the artery may be torn or one of the branches more often the anterior, or the veins which accompany the artery and its branches, and are placed nearer the bone. Sometimes the ascending branch is embedded in a channe in the bone and occasionally it pierces the squamous temporal bone and ramifies on its external surface beneath the temporal muscle. Hæmorrhage from these vessels may occur from head injury without fracture of the skull having been produced.

THE SPINAL CORD

Arteries

The spinal cord receives its blood supply from the anterior and posterior spinal arteries, the anastomosing sub-pial arterial network which connects them, and from the segmental arteries which join the anastomosis by entering the spinal canal along the issuing nerve roots. Intramedullary arteries penetrate the cord from the sub-pial anastomosis. The anterior, median placed, spinal artery which is the most important artery to the cord and the postero-lateral, usually paired, arterial trunks, arise from the vertebral; the segmental arteries arise from the vertebral, intercostal, lumbar and sacral arteries.

The blood supply of the cord is also largely dependent on from 6–8 anterior, and from 5–8 posterior radicular arteries, and from 6–11 anterior, and 5–10 posterior radicular veins. The largest of each set of vessels is found in the lumbar region as the arteria radicula magna and the vena radicula magna respectively. Every effort should be made to preserve the radicular vessels. Occasional instances in which section of a root has been followed by myelomalacia may have been due to the fact that the particular root divided has carried one of the main vessels. The middle thoracic part of the spinal cord has the poorest segmental circulation.

Veins

The veins of the cord are small, numerous and tortuous, but for the most part form longitudinal trunks which ramify on its anterior and posterior surfaces and in relation to each set of nerve-roots. The state of the veins which run longitudinally on the back of

Fractures of the base are usually associated with unconsciousness, which may last for some time, because of the violence which has been applied to the brain stem. There may be hæmorrhage or discharge of cerebrospinal fluid from the ears or nose as these fractures are frequently compound, opening into the subarachnoid space by way of these cavities. We would emphasize however that any case of head injury must be assessed on its clinical features which in the main are determined by the damage which has been done to the brain, its membranes and vessels and the reaction produced by their injury. The state of the bone is of somewhat secondary importance and indeed extensive comminution is sometimes a saving factor to the brain, inasmuch as the shattering of the bone has to a certain extent expended the force of the blow delivered to the skull and prevented its concentration on the contents at one particular site.

Fractures of the vault of the skull may be simple or compound, depressed or comminuted and may or may not be associated with damage to the brain and its membranous coverings and vessels. Intracranial hæmorrhage, which may be extra-dural or intra-dural and in the latter case extra-cerebral or intra-cerebral, may complicate a head injury. The most frequent form of traumatic intracranial hæmorrhage is subdural and extra-cerebral, the next extra-dural, usually from the meningeal vessels. These vessels may be torn, even when there is no fracture of the skull, though in practice most cases of meningeal hæmorrhage are associated with fractures crossing the line of the vessels as they lie between the dura and the bone or within the bone itself.

The soft skulls of babies may be indented without being fractured (Fig. 4, p. 14).

Concussion or Commotio Cerebri

This is usually regarded as a transient condition due to head injury in which there is immediate loss of consciousness, flaccid paralysis, amnesia for the event which caused it and recovery without any obvious aftermath. Of the many views advanced to account for the condition, a direct concussive effect on the brain cells (the neuronal hypothesis) is most widely held today.

Cerebral contusion and laceration usually result in loss of consciousness which is of longer duration than occurs in pure concussion and if such lesions occur in the brain stem, the stupor may be prolonged or terminate in death from medullary failure.

Cerebral Irritation

Cerebral irritation or traumatic delirium occurs in some cases, usually of closed head injury in which there has been contusion of the frontal or temporal poles. It is best treated by early depletion therapy but the subject of it may require paraldehyde or chloral formamide and in some cases small doses of hyoscine have been effective in controlling the noisy restlessness of these patients.

Compression of the brain such as may occur after a head injury results in slowing of the pulse and if the compression involves the motor area of the outer surface of one hemisphere, monoplegia or hemiplegia as well. The pupil on the side of the compression is dilated owing to the oculo-motor nerve being affected in its course across the gap in the tentorium. Parasympathetic fibres which innervate the ciliary body and the sphincter pupillæ lie in the upper part of the nerve and are vulnerable to pressure from above and laterally by the herniated uncus. Such pressure must also result in stretching the nerve.

tumour in the frontal region may not, if it has been slowly growing, affect the intracranial pressure to any extent.

The pressure of the cerebrospinal fluid rises during sleep as may be seen by observing the state of tension in a bony defect in the skull of a sleeping patient. This accounts for the fact that the headache of raised intracranial pressure is characteristically worse on the patient's waking from sleep.

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THE BRAIN AND SKULL

The Scalp

The scalp is a frequent site of sebaceous cysts which are easily extirpated and today complications such as horns and Cock's peculiar tumour are only very rarely seen. It is also a site of predilection for cirsoid aneurysms and these lesions are best treated by excision (Rogers, 1936, 1955).

Hæmatomas may form at a site of injury particularly in the scalps of children. These lesions rarely require treatment, the blood being absorbed in the course of time but when they are palpated by an inexperienced observer they may suggest the presence of a depressed fracture because the central area of effused blood is softer than the periphery.

INJURIES

Scalp Wounds

These may be punctured, incised, lacerated or contused and involve the scalp alone or the skull and its contents, in which case they are known as penetrating injuries.

Fractures of the Vault and Fractures of the Base of the Skull

In older textbooks sharp distinction is made between fractures of the vault and fractures of the base of the skull but it is doubtful whether any useful purpose is served by such distinction Fractures frequently extend from the vault into the base or vice versa, and while basal fractures usually imply gross cerebral damage and fractures of the vault may occur with little damage to the contents of the skull, this is not always the case.

Sedation

Aspirin, codeine phosphate and phenacetin, separately or in combination are useful for the relief of headache and sodium phenobarbitone (gr. 1½–3) may be given by injection if there is restlessness as well. Potassium bromide and chloral hydrate are also useful and can be given by mouth, through the stomach tube, or by rectum (twice the dose) if restlessness is severe; paraldehyde may be given by mouth, rectally or intramuscularly. Restlessness is less likely if cerebral œdema is checked by the early institution and maintenance of rectal magnesium sulphate infusions (vide supra).

If during the course of such treatment focal signs (such as monoplegia, hemiplegia or ophthalmoplegia) arise as epiphenomena these point to an expanding lesion due to the accumulation of blood and the necessity for exploration of the site indicated by the clinical picture.

INDICATIONS FOR OPERATIVE TREATMENT

The Posterior Fossa Compression Syndrome

Pressure by blood clot in the posterior fossa may cause respiratory failure while the heart continues to beat. Artificial respiration is carried out and as long as the respiratory exchange is maintained the patient's colour may be good and the pulse full and regular, but if the artificial respiration is not maintained the patient becomes cyanosed and the pulse slows and may stop, to start again with renewal of the artificial respiration. In such cases an airway should be introduced into the trachea and the posterior fossa opened without delay because if the pressure on the medulla can be reduced, the respiratory centre may begin to function again. It is useless to carry out prolonged artificial respiration in the hope that the natural respiratory rhythm will be established spontaneously.

Tracheotomy

If there is much cyanosis with respiratory difficulty, tracheotomy may be advisable as an emergency measure not only to improve the respiratory exchange but to facilitate aspiration of the bronchial tree.

Penetrating Injuries

If the injury is penetrating, i.e. a compound fracture of the skull, surgical toilet should be instituted as soon as the patient's condition permits, its object being as in the case of compound fractures occurring elsewhere in the body, to convert an open into a clean closed lesion. In dealing with these cases, particularly if the meninges have been lacerated and the brain penetrated, adequate facilities for major operative work should be available, such as skilled anæsthesia, endothermy, suction, supplies of blood and plasma, "gelfoam," fibrin film, etc., and the requisite special instruments for craniotomy and intracranial operations.

When operating for a compound fracture of the vault of the skull, the excision of the wound, because of the rich blood supply of the scalp, need be minimal only, an eighth of an inch or less being usually all that is required. There is no need to be radical in the removal of pieces of broken bone (Fig. 2). Fragments which are not obviously dirty or devitalized may be retained and should be so retained provided that the wound toilet can be carried out effectively. Torn dural edges should be trimmed and foreign bodies

THE TREATMENT OF HEAD INJURIES

Whenever possible the patient's skull should be X-rayed to ascertain the state of the bone in relation to the course of the meningeal vessels and venous sinuses and whether or not there is depression. Surgical shock is not a prominent feature of most head injuries but if present as a complication of associated injuries requires first consideration in the treatment of the patient.

There are comparatively few indications for early operation in cases of head injury. Scalp wounds should however be carefully excised and sutured as soon as the patient's condition permits, in order to prevent complications from infection. The object of treatment of the patient with a head injury is first the preservation of life by the restoration of lost blood, the appropriate treatment of concomitant injuries and resuscitation from shock, where such exists, and next the preservation of cerebral function by assisting the recovery of the damaged brain by maintaining its blood supply, the provision of rest for it and its freedom from compression. Intracranial venous congestion should be prevented by making sure that there is no constriction of the patient's neck by collars or bandages, while brain swelling, which may impair the blood supply and so lead to some degree of anoxia and cellular atrophy, is checked by limiting fluid intake and by introducing magnesium sulphate solution at intervals into the rectum (six ounces at 6-hourly intervals of a 30 per cent solution). The instillation must be slow to avoid rapid distension of the bowel and rejection of the solution and a watch should be kept on the blood urea concentration, particularly in children or if the administration has to be maintained for some time (e.g 10–20 days). As recovery takes place the rectal instillations are replaced by sodium sulphate given by the mouth. Although this method of depletion therapy is not universally approved we have found it advantageous and believe it is not only rational but that in some instances it is life-saving and that complications are reduced, if it is instituted early. It is interesting to note that during the late war several observers noted how much better cases of closed head injury did in the tropics than in more temperate climates and it is suggested that this may have been the result of a certain degree of dehydration.

Special care is needed in the case of the deeply unconscious patient who should be nursed semiprone with the head lowered, and turned at 2-hourly intervals. This allows the drainage of blood, cerebrospinal fluid, vomit or bronchial secretions from the pharynx and larger air passages and diminishes the risk of chest complications, indeed it may be a life-saving measure by preventing suffocation by inhaled fluid. If unconsciousness persists for any length of time it is necessary to attend to the fluid and nutritional requirements of the patient and this is best achieved by passing a small stomach tube (Ryle's tube) which may be left in position and removed every 48 hours for cleaning.

Lumbar puncture is best avoided in most head injuries particularly in the early phases. It can yield little information of value in assessing the case and may be harmful. After a few days or weeks, however, when there is headache and consciousness is still clouded, lumbar puncture every few days with the withdrawal of small quantities of fluid gives much relief to the patient and assists convalescence.

A common cause of restlessness in a semiconscious or unconscious patient is a full bladder and often much can be accomplished by intelligent anticipation on the part of the nursing staff.

Sedation

Aspirin, codeine phosphate and phenacetin, separately or in combination are useful for the relief of headache and sodium phenobarbitone (gr. 1½–3) may be given by injection if there is restlessness as well. Potassium bromide and chloral hydrate are also useful and can be given by mouth, through the stomach tube, or by rectum (twice the dose) if restlessness is severe; paraldehyde may be given by mouth, rectally or intramuscularly. Restlessness is less likely if cerebral œdema is checked by the early institution and maintenance of rectal magnesium sulphate infusions (vide supra).

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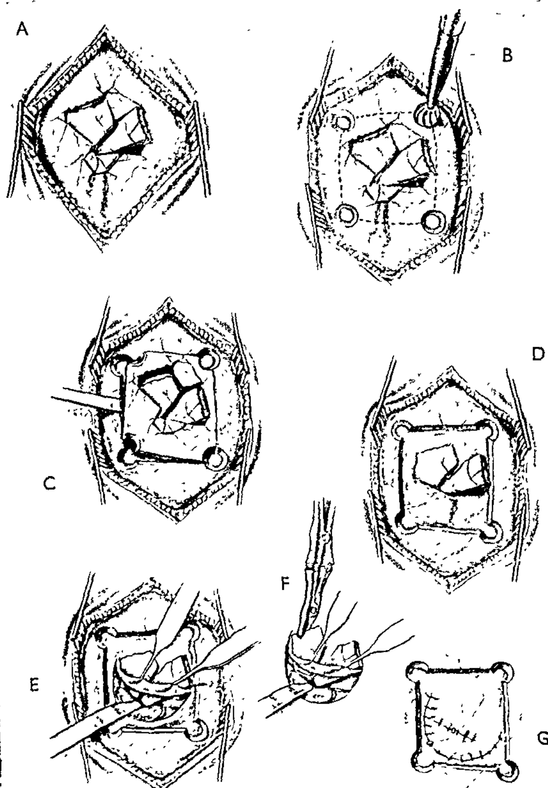
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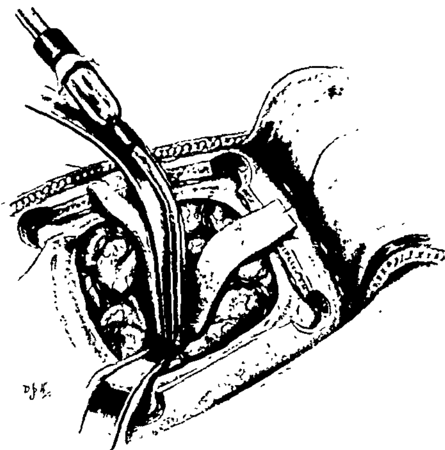
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(*From "Pictorial Introduction to Neurological Surgery" by G F Rowbotham and D P Hammersley, E & S Livingstone, Ltd*)

FIG 2 The treatment of compound depressed fracture

removed. Damaged tissue such as lacerated brain is removed by gentle suction combined with irrigation with warm saline or Ringer's solution (Fig. 3). In an early wound in which it has been possible to effect a satisfactory toilet the dura may be closed by using a fascial graft, but this is not essential and careful closure of the scalp in two layers is all that is necessary in an emergency. Any bony defect which remains is dealt with later. Drainage



(From "Pictorial Introduction to Neurological Surgery" by G. F. Rowbotham and D. P. Hammersley, E. & S. Livingstone, Ltd.)

Fig. 3. The use of suction for debridement of a wound tract.

is rarely necessary but if there is any doubt as to perfect hæmostasis or the state of the wound, a corrugated or small tubular, rubber drain should be placed in an appropriate part of the wound.

Closed Injuries

In closed or blunt injury the indications for operation are, elevation of depressed fractures (these are only rarely present, however, with an unbroken scalp and most depressed fractures are therefore compound injuries) and intracranial hæmorrhage, whether extra-dural or intra-dural or intra-cerebral. At a later stage, operative treatment may be required to deal with complications such as infection (abscess or osteomyelitis) and in cases of cerebrospinal rhinorrhea to close the opening in the meninges in order to prevent the spread of infection.

The indentations in the skulls of newly born babies (Fig. 4) sometimes disappear within a few weeks of birth but if they persist beyond this time they should be elevated by cutting and reflecting a flap of the bone in which the depression lies. This bone can be cut with a sharp pair of Mayo's or other suitable scissors. The indentation is then pressed out by the surgeon's thumbs and the flap replaced (Rogers, 1953).



(From "British Medical Journal")

FIG. 4 Baby boy aged 2 months with persistent depression in left frontal region

The Sequelæ of Cases of Head Injury

If care has been taken to check brain swelling the degree of recovery made by patients with even severe and extensive head injuries is remarkable. The former practice of keeping these patients for long periods in bed in darkened rooms has been superseded by getting them up early and by physiotherapy in the form of breathing and muscular exercises and massage. It is the practice of some surgeons to administer small doses of the barbiturates daily over long intervals such as 6–12 months or more, to patients who have had severe head injuries and to instruct their patients to avoid excitement, the use of alcohol and to limit or abolish smoking. These measures are taken with the object of lessening the incidence of sequelæ such as post-traumatic epilepsy, but it is difficult to determine just how effective they are.

After recovery from the early effects of an injury to the head a patient occasionally has persistent and sometimes troublesome headache. Lumbar punctures sometimes relieve this and if these alone are ineffective, air insufflation is worthy of trial. We have known it to be followed by complete and lasting relief.

Post-traumatic epilepsy may occur at varying intervals and is more liable to complicate cases in which the dura mater has been opened, the motor cortex involved, and in those in whom there is a family history of attacks of epilepsy or the epileptic equivalents. However, we have known a fit appear suddenly 3 years after an extra-dural hæmorrhage due to head injury in a young man in whom there was none of these criteria and despite all precautions; therefore a guarded prognosis must be given in most cases.

Bony Defects in the Skull

These are usually an aftermath of the treatment of compound fractures when the surgeon in carrying out the surgical toilet of the wound has removed pieces of the fractured vault of the skull. After sound healing has occurred it is usually desirable to

close these bony defects since otherwise they are sites of a weakened covering of the brain and so of possible injury to it. Furthermore they may have an ugly appearance, especially if in the frontal region, they may be a focus of interest or anxiety to the patient who realizes his skull is defective at the place where the opening lies and lastly if large the intracranial pressure may be affected. In the normal skull it is frequently slightly subatmospheric in the upright position as may be seen when cisternal puncture is performed, but if there is a large bony defect the pressure may be atmospheric. For all these reasons it is desirable to close these bony openings.

There can be no doubt that the correct material with which to close them is bone and this *can* be done no matter where they lie or how extensive they are. We would condemn the use of tantalum, vitallium, acrylic resin and all such substances which being foreign bodies may thereby direct the patient's attention to their presence, are opaque to X-rays and are not always well tolerated. In all these respects they have disadvantages over bone. If the opening in the skull is small it is closed with a piece of outer table from the nearby area of the vault; if larger, pieces of rib or ilium or tibia are used and if still larger and involving complicated contours such as large areas of the frontal bone, corresponding pieces of skull from the bone bank are most suitable. The results of all such reconstructive procedures are excellent provided only that a careful and highly aseptic technique is used. At the time of closure the adherent dura is separated from the bony edges and if the brain is adherent to the meninges this is first separated before closing any dural defect with fascia and filling in the bony defect in the manner indicated.

Injury to Cranial Nerves

Injuries to cranial nerves only rarely complicate head injuries. Ophthalmoplegia after injuries in the frontal region is as often due to direct injury to the ocular muscles as to their nerve supply. Anosmia may follow injuries to the cribriform plate and the nervous filaments from the olfactory lobes may be shorn off in contrecoup lesions. The anosmia is usually permanent and may be a great handicap in certain occupations. The optic nerve is sometimes damaged by a fracture through its foramen. Facial palsy after head injury is usually associated with fracture through the middle fossa and the same applies to the auditory nerve, injury to which is less common. In most cases the facial palsy recovers and no operation because of its apparent non-recovery should be considered until at least 6 months have elapsed.

Collet's Syndrome

This is the term given to traumatic lesions of the last four cranial nerves which may accompany injury to the floor of the posterior fossa. It is rarely seen but may result from falls either on to the head or the feet. As might be expected the syndrome consists of difficulty in swallowing, alteration of the voice, wasting of one half of the tongue and its deviation when protruded and weakness of the sterno-mastoid and trapezius muscles on the side of the lesion.

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Head Injuries

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The Scalp

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INTRACRANIAL HÆMORRHAGE

Bleeding inside the skull may occur in various anatomical situations and, apart from spontaneous subarachnoid hæmorrhage (vide infra), is usually associated with head injury. The types described in this section are (1) Extradural hæmorrhage, (2) Subdural hæmorrhage: (*a*) acute, (*b*) chronic, and (3) Intracerebral hæmorrhage.

Extradural or Middle Meningeal Hæmorrhage

This is due to damage to the meningeal vessels usually associated with a fracture of the skull, where quite often the fracture in the bone runs across the line of the vessels and lacerates them. Cases of meningeal hæmorrhage have occurred, however, without the skull being fractured (Rogers, 1937; Falconer and Schiller, 1942). The initial injury is often a slight one which may have produced only a short period of unconsciousness. After this the patient, as classically described, shows a "lucid interval" which may vary from less than an hour to 2–3 days. Then, with varying speed there is a progressive deterioration in the level of consciousness, leading eventually, in the absence of any intervention, to coma and death. This "classical" description does not conform to that most commonly seen, as there is often no definite lucid interval, and the only evidence of a progressive expanding intracranial lesion is a gradual deterioration in the level of consciousness sometimes associated with focal signs. This emphasizes the need for a thorough examination of the patient as soon as possible, with an accurate description of his conscious level recorded in the notes at successive examinations. The position of any bruises or lacerations in the scalp has been shown to be of value in the localization of the hæmatoma, but often the external injury may be comparatively slight and only apparent after the head has been shaved. The growth in the size of the hæmatoma is shown by the progress in the clinical signs and in the appearance of symptoms and signs of cerebral compression. A contralateral hemiparesis may develop and if the dominant hemisphere

is involved there may be aphasia. The progressive rise in pressure produces a tentorial pressure cone (Jefferson) with herniation of the uncus and pressure on the oculomotor nerve. The posterior cerebral artery may also be compressed or kinked, with the production of infarction of the occipital lobe and a homonymous hemianopia. The pressure on the oculomotor nerve may produce irritation in the early stages with a constricted pupil, but this rapidly progresses to a paralysis and the pupil gradually dilates and loses its response to light. The patient may only be seen at the stage when the pupils are dilating and inactive. This dilatation of the pupil is more advanced on the side of the hæmatoma and while it is helpful in showing the side of the lesion it also indicates the need for urgent intervention. If the tentorial herniation is allowed to progress there is likely to be serious pressure on the mid-brain with distortion and hæmorrhages, leading to a fatal result. Parallel with the changes in the level of consciousness there is quite often a fall in the pulse rate and a rise in blood pressure, but in the later stages when medullary failure is imminent the pulse becomes rapid and feeble.

When there is evidence of a progressive lesion as described, it is vital that treatment should be instituted without delay. Radiographs of the skull should be taken if delay is not entailed thereby, and these may produce helpful information by showing the position of fracture lines or a pineal shift. Lumbar puncture does not usually yield helpful information and as it may cause further delay and is not without risk, it is better avoided.

When the above features present there is usually no doubt that exploration is called for, although the exact diagnosis may not be certain. If a lucid interval occurs it is not entirely characteristic, as it has been seen in acute subdural hæmatomas. Exploratory burr holes in the fronto-parietal and occipito-parietal regions should never be omitted if there is any suspicion of an intracranial clot; a negative exploration causes little disturbance and is far better than missing a hæmatoma with possibly fatal consequences.

Treatment. In planning the position of exploratory burr holes the clinical picture of pupillary changes, hemiplegia or aphasia gives some guidance, but one should pay particular attention to the history and the nature of the accident, the position of scalp bruising and the site of the fracture lines (Lewin, 1949). From such information one is enabled to place the burr opening in such a position that the clot is more likely to be found and this is more satisfactory than an unconsidered, routine, bitemporal exploration.

As the patients are comatose or stuporose the burr holes can usually be made under local analgesia and the clot commonly extrudes spontaneously as the bone opening is made. There may be a definite single bleeding point to control or there may be a more diffuse venous ooze from the whole surface of the dura, particularly when the hæmatoma is extensive or has been present a long time. Bleeding points are usually easily controlled by coagulating diathermy or the application of gel-foam, muscle, or other of the hæmostatic agents. It is rarely necessary to ligature the main trunks of the meningeal vessels or their branches. Drainage is rarely necessary but if there is doubt about oozing, a small glove drain is inserted for 48 hours or longer as may be required. Occasionally hæmatomas occur in unusual sites such as far forwards in the frontal region or even on the floor of the anterior fossa, in the parietal region and further back in the posterior fossa, overlying the cerebellum. If care has been taken in the pre-operative assessment of the case these hæmatomas in unusual situations are less likely to be missed. However,

Rogers, Lambert (*a*) (1933) *Brit. med. J.* 2, 100.

Sydney, 3, 17.
1, 90.

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continual vigilance and often very speedy action are necessary in dealing with these cases and the existing high mortality presents a challenge to all surgeons. The need for prompt action often precludes the transfer of the patient to a special centre and all who handle accident cases should be prepared to treat these problems when they arise.

Acute Subdural Hæmatoma

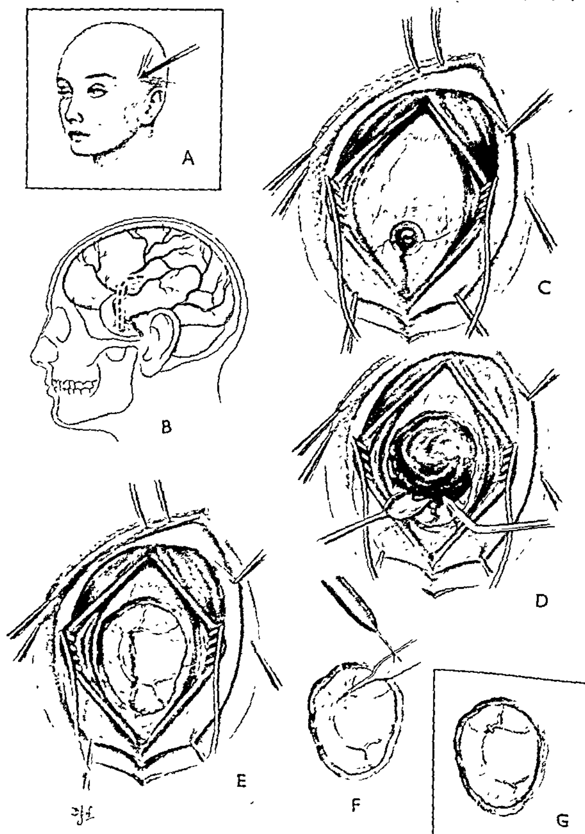
Acute subdural hæmatomas are usually the result of a more severe head injury than that associated with extradural hæmorrhage. The initial period of unconsciousness is usually longer, there may be no lucid interval, and only a progressive deepening of coma and advance in the physical signs betray the presence of an expanding lesion and indicates the need for exploration. Symptoms may be slower in manifesting themselves and may develop any time up to 14 days after injury. This is an arbitrary dividing line but a case with a longer interval should be regarded as chronic subdural hæmatoma (vide infra). As the bleeding often arises from laceration of the brain and not from torn veins alone there may be evidence of brain damage, while in some cases the bleeding is bilateral and the localizing signs may be misleading. Because of the associated brain damage the mortality is higher than in extradural clots; also in some cases there is an associated intracerebral clot, the recognition and treatment of which improves the chances of recovery.

The treatment is essentially the same as that for extradural clots and consists of making exploratory burr openings and washing out the clot. If the clot is more solid or if there is bleeding which cannot be controlled it may be necessary to turn a small osteoplastic flap. The burr holes should always be planned with this possibility in mind so that if the need arises they can be incorporated in the flap. The same remarks apply to the question of drainage as have been made in the section on extradural hæmorrhage.

Chronic Subdural Hæmatoma

Ætiology. Although injury is a factor in the production of this condition it is often quite slight and may have occurred as long as 12 or more weeks beforehand, but in roughly one-third of cases there is no evidence at all of earlier head injury. This seems to be particularly the case in the older subjects and it may be that a low intracranial pressure and a shrunken brain with large subdural space traversed by unsupported veins, together with abnormal vascular fragility and dehydration, predispose to the condition Under these circumstances a very minor strain or blow may cause rupture of a vein with a slow leakage of blood which eventually collects as a subdural hæmatoma. Cases have been reported in which the head injury was "indirect" and produced by a heavy fall on the buttocks. In many instances it seems that those cases with the longer history are those in which the injury was minimal Once the original hæmatoma has collected and formed an organized membrane, further growth is possible by osmosis even if all bleeding has stopped. Thus some collections consist mainly of fluid, varying from deep yellow to dark red in colour whilst in other cases there is much more solid blood clot, or else altered blood forming a yellowish-brown sludge inside the membrane, with or without the fluid component.

Many of the patients, particularly those in whom the history of injury is absent, are over 50, but the condition also occurs in younger people and we have seen one example in a boy aged 9, where there was a clear history of minor head injury 8 weeks earlier.



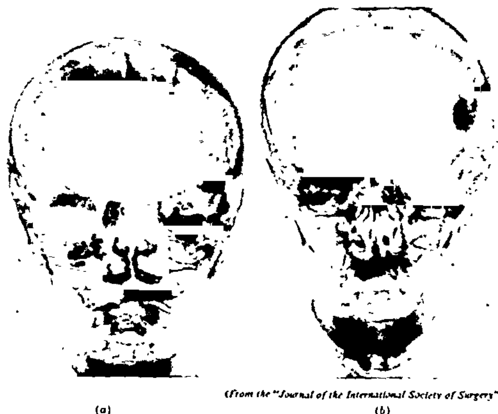
(From "Pictorial Introduction to Neurological Surgery" by G. F. Rowbotham and D. P. Hammersley, E. & S. Livingstone, Ltd.)

FIG. 5. Steps in the treatment of extradural (middle meningeal) hæmorrhage

Straight films of the skull may show no characteristic change but in unilateral hæmatomas a shift of a calcified pineal, which is displaced downwards as well as to the opposite side, may be revealing; and rarely, calcified lesions have been described.

The electroencephalogram is not characteristic but is abnormal in a high percentage of cases and there may be a "silent" area with suppression of the normal electrical activity under the hæmatoma or an irregular continuous slow activity of low or moderate voltage (Rogers, 1941).

Angiography is of great value and gives a characteristic picture in a large proportion of cases. It may be most obvious in the capillary phase ("first phlebogram") when there



(*From the "Journal of the International Society of Surgery"*)

(*a*) (*b*)

FIG. 7 (*a*) Subdural air filled cavity from which a hæmatoma had been removed in a man aged 62 who had fallen off his bicycle and knocked his head three months before the operation for evacuation of the blood clot. (*b*) The same patient three months later. The cortex has come up to obliterate the space.

is a lenticular area between the skull and the brain which shows no vessels (Fig. 6). In the arterial phase the branches of the middle cerebral artery are displaced medially and there is also a displacement of the anterior cerebral artery to the opposite side.

Pneumography is not often necessary in making a diagnosis and if burr holes are being made in a case of suspected intracranial tumour it is quite common to encounter the hæmatoma and drain it, without the necessity of injecting air. If the ventricles are needled the pressure of the contained fluid is usually low. There is also low pressure in the subdural space, and after the effusion has been drained the space may take some time to become obliterated. It may fill with air which outlines its extent and this may show in suitable radiographs (Fig 7 (*a*), (*b*)).

Treatment consists of making one or two burr holes on each side of the head, draining

The condition is also met in the neonatal period when it is probably the result of birth injury.

Diagnosis. There is often a history of persistent headache after a head injury, but in some cases there is an insidious or rapid onset of raised intracranial pressure, which may mimic a rapidly growing glioma. The severity of the headache may fluctuate considerably but often there are very severe paroxysms which may be accompanied by vomiting and this may be the main reason for the patient seeking treatment. As the condition progresses



Fig 6 Angiogram of subdural hæmatoma in woman of 69, showing characteristic appearance, with shift and filling defect

there is increasing drowsiness although this may show great variability from day to day, or even from hour to hour. Parallel with this there may be mental changes varying from a mild degree of confusion to quite severe dementia, which may result in the patient's admission to a mental hospital. In the past it is probable that many patients with these hæmatomas died undiagnosed in asylums, or were regarded as cases of "pachymeningitis hæmorrhagica" which was frequently described as one of the post-mortem findings in the insane. Less commonly there may be one or more major epileptic attacks and in some cases slight changes in personality may be all that is apparent

Lumbar puncture shows quite a low cerebrospinal fluid pressure in some cases although in others it is normal or raised. In long-standing cases, the fluid may be yellow and the protein content raised.

puncture in order to encourage the expansion of the brain and perhaps to disimpact a tentorial pressure cone.

A subdural hæmatoma sometimes develops as the result of rupture of an intracranial aneurysm, usually one on the internal carotid artery, where it is in relation to the basal arachnoid cisterns. In these cases the arachnoid may be torn and the blood in addition to producing the expected subarachnoid hæmorrhage burst through into the subdural space. As the subarachnoid hæmorrhage dominates the clinical picture it may be difficult to diagnose the separate existence of a subdural effusion, but this may be quite large and the advantage in such a case of a direct attack on the responsible aneurysm is apparent as the hæmatoma can be treated at the same time.

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SPONTANEOUS (NON-TRAUMATIC) INTRACRANIAL HÆMORRHAGE

(1) Subarachnoid

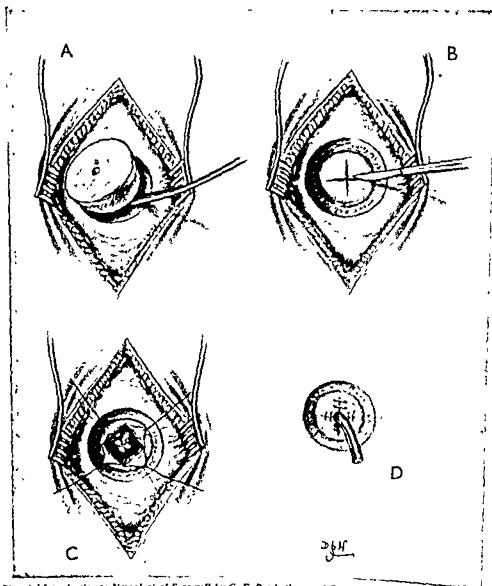
(2) Intracerebral

Spontaneous Subarachnoid Hæmorrhage

> And when the child was grown, it fell on a day that he went out to his father to the reapers. And he said unto his father, My head, My head. And he said to a lad, Carry him to his mother. And when he had taken him, and brought him to his mother, he sat on her knees till noon, and then died.
>
> 2 Kings, 4, 18–20.

Spontaneous subarachnoid hæmorrhage is one of the commoner causes of sudden death or invalidism in young people. It may come as a bolt out of the blue. Simpson (1947) records the case of a young man who worked in a garage. He was lying under a car tightening a nut with a spanner when he suddenly cried, "Oh my head," collapsed and was dead when a doctor reached him only 4 or 5 minutes later. It is not always as catastrophic and a case has been recorded in a fighter pilot who despite the fact that he had a hæmorrhage when airborne successfully landed his aircraft (Rash and Goldys, 1945). One of our patients was driving a van when he had an intense headache, drove his van into the side of the road and stopped the engine. He was taken out of the van unconscious and subsequently made a good recovery from a subarachnoid hæmorrhage. While headache, usually of sudden onset, is almost invariable not all patients lose consciousness. The rate and amount of the bleeding may vary from a trickle to a torrent. The most frequent cause of subarachnoid hæmorrhage is the rupture of an intracranial aneurysm (Fig. 9 (*a*), (*b*) and Fig. 10) either on the circle of Willis, the intracranial portion of the internal carotid artery, the vertebral or basilar artery or one of their branches.

away the hæmatoma fluid, and irrigating the subdural space with Ringer's solution (Fig. 8). In exceptional cases the contents of the hæmatoma may be firmly clotted and too thick to drain through burr holes and it will then be necessary to turn an osteoplastic flap. In some cases the flap may be turned because a tumour is suspected, and the finding of a



(From "Pictorial Introduction to Neurological Surgery" by G F Rowbotham and D P Hammersley, E & S Livingstone, Ltd)

Fig 8 The treatment of subdural hæmatoma In the illustration a trephine has been used but burrs are more often employed

hæmatoma is an agreeable surprise. In an adult it is not essential to remove all the membrane, because it will not interfere with the development of the cerebrum but it is conceivable that it might do so in an infant with a subdural hæmatoma and in such it is better removed.

Because of the low pressure frequently found, and the necessity of obliterating the hæmatoma cavity as soon as possible, these patients are nursed with the head low, the foot of the bed raised and a generous fluid intake is encouraged. In rare instances it is advisable to drain the cavity and inject saline into the subarachnoid space by lumbar

puncture in order to encourage the expansion of the brain and perhaps to disimpact a tentorial pressure cone.

A subdural hæmatoma sometimes develops as the result of rupture of an intracranial aneurysm, usually one on the internal carotid artery, where it is in relation to the basal arachnoid cisterns. In these cases the arachnoid may be torn and the blood in addition to producing the expected subarachnoid hæmorrhage burst through into the subdural space. As the subarachnoid hæmorrhage dominates the clinical picture it may be difficult to diagnose the separate existence of a subdural effusion, but this may be quite large and the advantage in such a case of a direct attack on the responsible aneurysm is apparent as the hæmatoma can be treated at the same time.

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SPONTANEOUS (NON-TRAUMATIC) INTRACRANIAL HÆMORRHAGE

(1) Subarachnoid

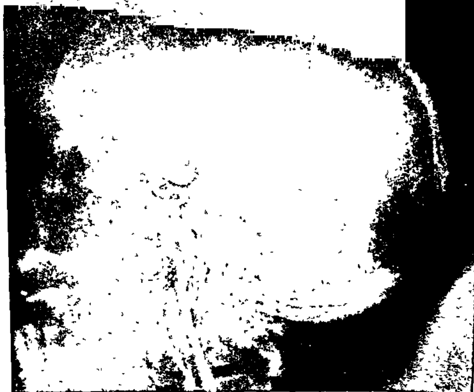
(2) Intracerebral

Spontaneous Subarachnoid Hæmorrhage

> And when the child was grown, it fell on a day that he went out to his father to the reapers. And he said unto his father, My head, My head. And he said to a lad, Carry him to his mother. And when he had taken him, and brought him to his mother, he sat on her knees till noon, and then died.
>
> 2 Kings, 4, 18-20.

Spontaneous *subarachnoid hæmorrhage is one of the commoner causes of sudden* death or invalidism in young people. It may come as a bolt out of the blue. Simpson (1947) records the case of a young man who worked in a garage. He was lying under a car tightening a nut with a spanner when he suddenly cried, "Oh my head," collapsed and was dead when a doctor reached him only 4 or 5 minutes later. It is not always as catastrophic and a case has been recorded in a fighter pilot who despite the fact that he had a hæmorrhage when airborne successfully landed his aircraft (Rash and Goldys, 1945). One of our patients was driving a van when he had an intense headache, drove his van into the side of the road and stopped the engine. He was taken out of the van unconscious and subsequently made a good recovery from a subarachnoid hæmorrhage. While headache, usually of sudden onset, is almost invariable not all patients lose consciousness. The rate and amount of the bleeding may vary from a trickle to a torrent. The most frequent cause of subarachnoid hæmorrhage is the rupture of an intracranial aneurysm (Fig. 9 (*a*), (*b*) and Fig. 10) either on the circle of Willis, the intracranial portion of the internal carotid artery, the vertebral or basilar artery or one of their branches.

(*a*)



(*b*)



FIG. 9. Aneurysm on internal carotid artery which produced oculomotor palsy and also subarachnoid hæmorrhage.

Occasionally bleeding arises from an intracranial angiomatous malformation (arterio-venous aneurysm, Fig. 11 (*a*), (*b*)) and more rarely from a tumour (Echols and Rehfeldt, 1950), a mycotic aneurysm, or in association with certain blood dyscrasias.

Aneurysms occur much more commonly on the anterior half of the circle of Willis but they are also found on the vertebral, basilar and posterior cerebral arteries.



FIG. 10 Calcification outlining wall of large intracranial aneurysm in a woman aged 55. A similar appearance may be produced by some supra-sellar cysts (cranio-pharyngiomas) and some meningiomas

Ætiology and Pathology

Berry aneurysms occur on the cerebral arteries of from 0·25–1·5 per cent of subjects (Glynn, 1940). Various explanations have been advanced to account for their development. Probably they are congenital in origin as suggested in 1887 by Eppinger and are survivals of the embryonic vascular net-work (Bremer, 1943). That they may not cause trouble until late in life is due to secondary factors such as changes in the vessel wall, either defects in the medial, muscular coat, or in the elastic layer. Such defects seem particularly liable to occur at the points of bifurcation of the cerebral vessels, and it is in these situations that a very large number of these aneurysms are found.

Complications

An intracranial berry aneurysm may rupture at any time and neither strain nor trauma appear to play any significant part, as rupture may occur when the patient is at rest, or even when in bed asleep. Bleeding may be torrential and rapidly fatal or merely a slow leakage. A small escape of blood causes symptoms of headache, giddiness and pain in the neck and occasionally when blood accumulates in the spinal subarachnoid space there may be severe pain in the back and legs due to irritation of the spinal roots.

If the aneurysm is on the circle of Willis in the basal cisterns the bleeding occurs freely into the subarachnoid space but if it is on the middle cerebral artery in the Sylvian

(a)



(b)

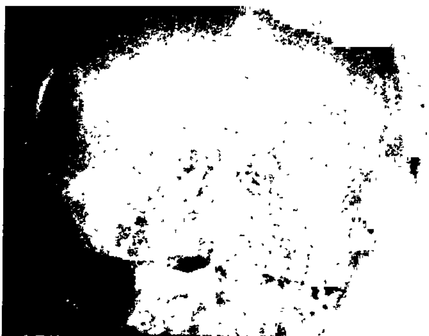


FIG. 9 Aneurysm on internal carotid artery which produced oculomotor palsy and also subarachnoid hæmorrhage.

fissure, or on the anterior cerebral or anterior communicating arteries, the blood may tear its way into the brain substance. This has been noted in cases where an earlier hæmorrhage has caused adhesions and blocked the free passage for the blood into the subarachnoid space. Successive small intracerebral hæmorrhages of this description cause areas of softening which may result in a final hæmorrhage bursting through into the lateral ventricles.

Occasionally thrombosis occurs in an aneurysmal sac, particularly the larger saccular lesions and this may result in a spontaneous cure. Such thrombosis has also been noted after proximal ligation of the carotid artery, performed as a curative measure (Figs. 12, 13, 14, 15). The site of rupture of the sac of an aneurysm varies. In many instances the fundus gives way but sometimes the weakest place appears to be the neck of the aneurysm at its junction with the parent artery (Small *et al.*, 1953).

Symptoms and Signs

Some of these have already been described. In a typical case there is a sudden onset of severe headache, which may be felt in the frontal region and even over the face, or it may be vertical and spread to the back of the head and into the neck. There may be vomiting and collapse but consciousness is not always lost. Later, stiffness of the neck develops and there may be other symptoms due to pressure on neighbouring structures. Thus there may be weakness or paralysis of limbs, squints or ptosis and pain in the face or frontal region. Unilateral frontal pain and ptosis localize the lesion to the carotid territory of that side.

A hemiparesis if present is not always of localizing value as an aneurysm arising from one anterior cerebral artery may rupture into the opposite frontal lobe and produce ipsilateral signs. The patient may be deeply unconscious, stuporous and confused or may be conscious and co-operative. There may be mental deterioration and a grasp reflex and if the dominant hemisphere is involved, dysphasia also. An isolated third nerve palsy is most commonly caused by an aneurysm on the posterior communicating artery (Jefferson, 1947) which may not have ruptured. Examination of the fundus oculi may show no abnormality in the early stages, but there may be slight blurring of the optic disk, more marked swelling with hæmorrhages or prominent subhyaloid hæmorrhages. If these are large or in the macular region they produce prominent scotomata or even complete blindness in this eye. These changes result probably from stasis in the orbital and ophthalmic veins. It is unusual to find subhyaloid hæmorrhages other than in a first attack because the extension of the subarachnoid space along the sheath of the optic nerve, which is the pathway for the spread of the extravasated blood, becomes obliterated by adhesions and prevents a recurrence.

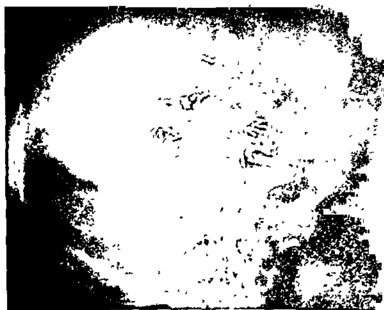
Bleeding into the hemisphere may be suspected if there is a more prolonged period of unconsciousness or if there are localizing signs such as hemiparesis, aphasia or hemianopia. Occasionally in these cases there is little or no subarachnoid bleeding and the appearance of the cerebrospinal fluid may be misleading.

Investigations

The diagnosis is confirmed by lumbar puncture which reveals a blood-stained cerebrospinal fluid which may show all variations from a slight pink colour to a fluid resembling pure blood. The state of the fluid does not always reflect the severity of the hæmorrhage,



(a)



(b)

FIG 11 (a), (b). Angiomatous malformation in man of 48 First film shows "tumour" and abnormal arterio-venous shunt The postoperative film shows the great improvement in the circulation through the normal cerebral vessels

as in those with a lightly stained fluid the bleeding is sometimes predominantly intracerebral. The pressure of the fluid may be raised, in some instances to more than 300 mm. When the cells are allowed to settle the supernatent fluid is pink or yellowish in colour.

Plain radiography may sometimes demonstrate abnormalities in cases of aneurysm. There may be calcification in the sac wall or erosion of bone or a pineal shift if there is a large hæmatoma. Angiography provides the most useful information for determining the subsequent management of the patient and should be done as soon as the patient's condition permits. This investigation in most cases confirms the diagnosis and reveals the size and site of the lesion. It should always be bilateral as localizing signs may be misleading and intracranial aneurysms are sometimes multiple. It also provides an opportunity to test the adequacy of the cross-circulation and enables the type of surgical attack to be planned. Angiography also enables us to exclude tumour or angioma as a cause of the symptoms and it may demonstrate intracerebral or subdural clot, the existence of which might affect the management of the case. A "complete" angiographic investigation should include injection of the vertebral artery but this may not always be expedient.

In certain cases air encephalography or ventriculography may be of value in revealing intracerebral or intraventricular clot.

Matas's test of digital compression of the carotid in the neck provides a useful indication of the patient's ability to tolerate carotid occlusion and in some instances where the cross-circulation has appeared inadequate, it has been improved by accustoming the patient to progressively longer occlusion of the artery (Rogers *et al.*, 1953).

Treatment

If the hæmorrhage is not rapidly fatal and investigations such as have been outlined above have revealed the nature and site of the lesion treatment is indicated either to arrest continued bleeding or to guard the patient against further hæmorrhages in the future. This treatment may be either by:

INDIRECT ATTACK—CAROTID LIGATION.
DIRECT ATTACK—LIGATION, CLIPPING, REINFORCEMENT.

Carotid Ligation

Ligature and division of the common carotid artery is of particular value in the treatment of aneurysms which lie on the internal carotid artery below the circle of Willis but it may also be used to treat aneurysms on the circle itself or even on its more peripheral branches. It has the merit of being easy to perform and its effectiveness is shown by the clinical improvement of the patients, by the evidence of angiography and by operative exposure of the treated aneurysm. It has been shown experimentally by Bakay and Sweet (1952), that there is also a reduction in pressure in the arteries above the circle after carotid ligation. A certain pressure has to be maintained through the collateral circulation however in order to prevent a hemiplegia developing through reducing the blood supply to one cerebral hemisphere and for this reason some surgeons favour treating these more peripherally placed lesions by direct exposure (v infra).

Before we embark on carotid ligation the patient's reaction to carotid occlusion will have already been tested (Matas's test), the only exception being in cases where the



Fig 12



Fig 13

Figs 12 and 13 Antero-posterior and lateral angiograms made in case shown in Fig 10, woman aged 55 with large aneurysm

(From "The Medical Press")

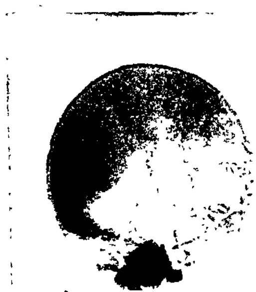


Fig 14



Fig 15

Figs 14 and 15. Angiograms in same case as in Figs 10, 12 and 13, 3½ years after division of the left common carotid artery The shrinkage in the size of the aneurysm is apparent.

(From "The Medical Press")

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operation is performed as an emergency to arrest continuing hæmorrhage. The operation is done under local anæsthesia so that when possible the full co-operation of the patient can be obtained and function in the opposite limbs constantly tested. The artery is occluded for 20–30 minutes and if no untoward symptoms develop it is then divided between ligatures (Fig. 16). This severance of the artery has the effect of producing a sympathetic denervation, encouraging the development of the collateral circulation and preventing subsequent embolic complications and we regard it as most important (Rogers, 1944, 1947, 1952, Lewis *et al.*, 1953). In some cases it may be necessary at a later date to ligate the internal carotid artery, particularly if there has been little or no improvement, e.g. persistence of oculomotor paralysis. The artery should be resected (Rogers, 1947).*

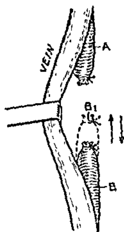


Fig. 16 To show how the ends of the divided common carotid retract and the pulsation in the proximal end as compared with the quiet, distal end

(From the "Lancet")

Some surgeons have practised occlusion of the artery in stages by ligating it with fascia, while others have used a clamp which is slowly occluded, such as the ingenious one devised by Selverstone of Boston. Poppen (1951) recommends a procaine block of the superior cervical ganglion to minimize spasm in the cerebral vessels, while others advocate anticoagulant therapy and papaverine injections, all with the object of maintaining adequate circulation in the hemisphere on the side of the ligation.

We emphasize however the need to divide the common carotid and where necessary to resect the internal carotid, believing that many of the complications of carotid ligation which have been reported can be ascribed to tying these arteries in continuity.

Direct Surgical Attack

A decision on this method of treatment will depend on the results of angiography, with regard to the position and relations of the aneurysm and the efficiency of the intracranial circulation. The advantage of a direct attack on the aneurysm is that the intracranial circulation may usually be left intact while there is also an opportunity at the same time to clear out any subdural or intracerebral clot that may be present as a result of earlier rupture of the aneurysm. Such a clot may already have been demonstrated by angiography and this would obviously tip the balance in favour of direct attack.

Aneurysms on the internal carotid, middle cerebral or anterior cerebral and anterior communicating arteries can be exposed by the routine frontal (Trotter or Naffziger) flap and it has been found convenient and satisfactory to use an extradural approach as in the exposure of the pituitary. The operations are now made easier and safer by the use of arterial hypotension (e.g. with "Arfonad").

When the aneurysm is exposed it may be treated in one of several ways. If it is clearly defined in relation to the surrounding vessels and is pedunculated, i.e. has a good "neck," it may be possible to ligature this with silk or occlude it with a silver or tantalum clip. Sometimes the shape or position of the sac may not permit this and it may then be obliterated by a larger clip applied directly or tangentially to the sac. Such treatment is accorded to aneurysms lying in the bifurcation of branches of the middle cerebral artery. Occasionally the sac itself is better "trapped" by occluding the parent vessel on each

* We have now performed over 100 carotid ligations.

side of it as may be done on the anterior cerebral, or by a ligature on the internal carotid in the neck and a clip on the carotid above the aneurysm, intracranially. It may occasionally be justifiable to sacrifice a small branch of the middle cerebral but care must be taken to spare the main trunk. Some aneurysms may not be ligated because of possible damage to perforating arteries or because of vessels passing out of them to supply important areas beyond. In cases where it is not possible to apply a clip or ligature, or also in some cases where this has been done, the wall of the aneurysm may be reinforced by a layer of hammered muscle, by a wisp of cotton wool or a sheet of cellophane or other plastic. By such means it is hoped to lessen the possibility of rupture in the future. Occasionally it may be possible to open the sac and pack it with muscle, after which it is ligatured or shrivelled up with the endothermy, so that the cavity is completely obliterated.

If an intracerebral clot is encountered it is cleared out and hæmostasis secured in the cavity. In middle cerebral aneurysms the clot is commonly in the temporal lobe while it is more commonly in the frontal lobe with aneurysms of the anterior cerebral or anterior communicating.

Intracranial Aneurysms without Rupture

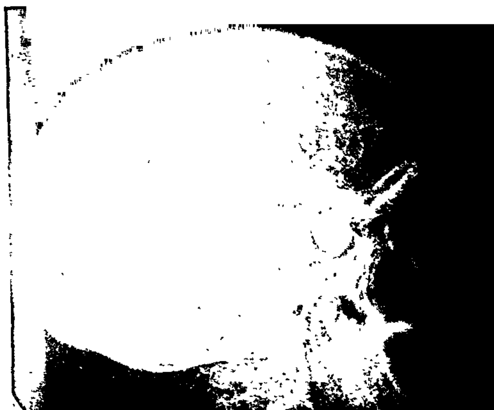
"Paralytic" and Fistulous Aneurysms

Not all aneurysms present with a subarachnoid hæmorrhage. Many, particularly those designated "infraclinoid" produce symptoms by pressure on neighbouring structures, more particularly the cranial nerves, but occasionally on the brain itself. They occur particularly in middle-aged women and commonly develop on that portion of the internal carotid which is in the cavernous sinus and as they enlarge the nerves in its wall are compressed Unilateral facial pain may be the sole complaint and a diagnosis of migraine may be made (ophthalmoplegic migraine). The aneurysm may show a sudden increase in "activity" with considerable increase in the pain. There may be at these times an associated progress in the clinical findings. Pressure on the different divisions of the trigeminal nerve causes numbness over the face and involvement of the various ocular-motor nerves produces varying degrees of ophthalmoplegia (Jefferson, 1938). There is often bony erosion of the sella tursica or sphenoidal fissure, which can be demonstrated in skull radiographs. The angiographic appearance may be quite striking (Fig. 17 (*a*), (*b*)) Upward and forward extension of the aneurysm may involve the optic nerve but this is more commonly pressed on by large aneurysms arising from above. There may be pressure on one optic nerve, or a more typical chiasmal syndrome may be caused. A large supraclinoid aneurysm in the frontal lobe may resemble a tumour (Fig. 10).

Carotico-cavernous Fistulous Aneurysms

In this condition the carotid artery, in its course through the cavernous sinus ruptures, so that there is a fistulous communication between the arterial and venous circulations. This means that the arterial pressure is transmitted to the local venous system and this accounts for most of the ill effects produced by the condition.

It may be caused by trauma associated with fractures of the skull base or arise spontaneously. In the former case the patient may be of any age or sex, but in the latter it most commonly occurs in middle-aged women in some of whom there is already an



(a)



(b)

Fig. 17. (a) Infraclinoid aneurysm in woman of 59 causing ophthalmoplegia and trigeminal anæsthesia. Successfully treated by ligation of common carotid artery. (b) The A.P. projection shows a similar lesion in a woman of 70—also successfully treated by carotid ligation.

aneurysm present in the sinus. Thus the rupture of the artery may be due to some weakness in its wall or an aneurysm which has caused no previous symptoms. There is an embryonic arterial communication between the carotid and basilar arteries ("trigeminal artery") and it is possible that in some cases a remnant of this forms an aneurysm and determines the site of rupture. In rare cases this carotid-basilar anastomosis persists into adult life, and since angiography has become a frequent investigation it has been demonstrated on a number of occasions during life.

There are rarely symptoms before rupture but occasionally the patient complains of headache of a migrainous type. In the traumatic cases rupture may either be immediate or delayed. When it occurs a bruit develops and may be the first thing noted by the patient. It is synchronous with the pulse and may be heard on auscultation over the frontal and temporal regions and over the carotid artery in the neck, and is usually maximal over the globe of the eye. It can be obliterated or greatly reduced by digital occlusion of the carotid artery in the neck. It is often quite loud with an echoing, reverberating character which is quite typical. It may be continuous with a systolic intensification.

There may be pain in the eye or forehead but this is by no means constant. As the arterial pressure is transmitted into the ophthalmic veins, exophthalmos develops but its rate of development is very variable. There is congestion of the scleral vessels and marked œdema of the conjunctiva develops. The condition is often described as "pulsating exophthalmos" but the pulsation of the eyeball may be difficult to detect on account of chemosis and swelling of the eyelids and on occasions it may be completely absent. The opposite eye may also be affected, but the extent of its involvement depends on the connexions between the two cavernous sinuses across the midline. There may be retinal hæmorrhages and venous congestion and subsequently a consecutive optic atrophy may develop with considerable failure of vision. Vision may be further affected by involvement of the cornea, when the degree of proptosis is excessive and ulceration occurs.

The treatment of this condition is ligation of the common carotid artery which should be divided between ligatures. This will produce a cure in about two-thirds of the cases. In the remainder the internal carotid may need to be occluded also and if so the artery should be resected and not just tied in continuity. Rarely it is necessary in addition to occlude it intracranially above the cavernous sinus. Before the carotid artery is permanently occluded the same precautions as have previously been noted should be observed to ensure that the cerebral circulation is adequate.

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Eppinger, H. (1887) [illegible]
G[illegible]
Je[illegible]
Je[illegible] 9.
Lewis, C., Langmaid, C. and Rogers, Lambert [illegible]
[illegible]



(a)



(b)

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that seen in cerebral abscess. Radiography may show change in the skull bones if the infection has been present for some time and there is definite osteomyelitis present (Fig. 19).



FIG. 18 (*a*) Pott's puffy tumour in a boy aged 16.
(*From "British Journal of Surgery," John Wright & Sons, Ltd.*)



FIG 18 (*b*). Pott's puffy tumour in a boy aged 11.
(*From "British Journal of Surgery," John Wright & Sons, Ltd.*)

Treatment

When there is suppuration in the scalp this is treated by incision and drainage. The incision in the scalp provides the chance to examine the skull for signs of infection. A burr opening should then be made to drain any extradural collection of pus. If no pus is found the dura should not be stripped as this may encourage the further spread of infection. Local instillation of penicillin may be an advantage and a rubber tube can be left *in situ* for regular irrigation.

SUBDURAL ABSCESS (Purulent Pachymeningitis)

*This condition is usually secondary to pansinusitis, associated with spreading osteo-*myelitis of the skull, mastoiditis, or blood-borne spread from more remote sites and

Rogers, Lambert (1949) *Lancet*, 1, 949.
Rogers, Lambert (1952) *Bull. Post. Grad. Comte. in Medicine*, University, Sydney, 8, 65.
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Thrombosis of the Internal Carotid Artery

Intermittent, unilateral attacks of impaired movement or sensation sometimes accompanied by disordered speech or vision in the eye of the same side may suggest the presence of an intracranial tumour but in reality be due to carotid thrombosis. In the latter however convulsions, alterations of consciousness and signs of raised intracranial pressure, typical of an expanding lesion, do not occur.

Carotid thrombosis affects men 3 times as often as women and the left internal artery is involved 6 times more often than the right (Perlow *et al.*, 1955). It may result from injury, particularly boxing, and we have seen examples in young men, but it may also arise spontaneously in young women or be the result of thrombo-angiitis or atherosclerosis somewhat later in life. The thrombosis is usually limited to a segment of the artery. Diagnosis is established by angiography. In the earliest phases anticoagulants are indicated. Resection of the thrombosed segment and sympathectomy in the later stages have not proved of much value.

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INFECTIONS

Extradural Abscess

This is usually associated with osteomyelitis of the skull which arises as the result of a direct spread of the infection from a frontal sinusitis or from infection in the mastoid region. It may also arise as a direct spread of infection through diploic veins or, more rarely, as a metastatic blood-borne infection from elsewhere in the body. Another occasional cause is direct injury associated with penetrating wounds or compound fractures. When there is osteomyelitis of the skull there is often an overlying cellulitis of the scalp giving rise to the swelling known as "Pott's Puffy Tumour" (*Fig 18 (a), (b)*). There may be superficial suppuration as well as that between dura and bone. On account of the adherence of the dura the abscess is usually well localized but if untreated the infection may spread and complications arise such as leptomeningitis, subdural or intracerebral abscess.

The commonest organisms are staphylococcus aureus, streptococcus (both anærobic and ærobic) pneumococcus and less commonly streptococcus viridans and B. proteus

The condition may be suspected when swelling of the scalp develops or when there is increased pain in association with a frontal sinus or mastoid infection. In the absence of intracranial extension of the infection there are not usually signs of neurological disorder. The cerebrospinal fluid may be quite normal, but on the other hand may resemble

the cerebellum the capsule may be quite well developed when the case first presents, and this has a bearing on the subsequent management. At a later stage the surrounding œdema may subside and the encapsulated abscess behaves like a tumour, as a "space-occupying lesion."

Ætiology. The infecting organisms reach the brain either by (*a*) direct spread or (*b*) through the blood stream.

(*a*) Direct spread occurs most commonly from the mastoid, and accounts for 50-60 per cent of all cases. From this site it may spread either upwards to the temporal lobe or backwards to the cerebellum. Spread from the frontal or other nasal sinuses accounts for another 7 per cent of cases. Other causes are osteomyelitis of the skull, penetrating wounds, including compound fractures and infections of the face, scalp, and skull.

(*b*) Hæmatogenous spread occurs most commonly from chronic chest infections such as bronchiectasis, empyema or lung abscess. The intracranial abscess is multiple in about half of the cases and occurs most commonly in the territory of the middle cerebral artery. Blood-borne infection may also originate in the abdomen or from boils and carbuncles, from infective endocarditis and occasionally from the infected mastoid. Such indirect spread from the mastoid, and also the more remote spread that sometimes occurs from the frontal sinus may occasion difficulty in diagnosis and localization.

Bacteriology. The organisms most commonly found are the Staphylococcus aureus, streptococcus and the pneumococcus, but S. viridans, B. proteus and M. tuberculosis may be responsible. In thoracogenic abscesses we find anærobic streptococci with or without fusiform bacilli, pneumococci, staphylococci and H. influenza. Less commonly seen are the actinomyces, diphtheroids and gram-positive and gram-negative rods. It is important to ascertain the causative organism if this is at all possible so that the appropriate antibiotic may be used in treatment. The preliminary treatment which a patient has received before any pus is aspirated may make organisms very difficult to culture, even though they have not been destroyed and have been seen in stained films of the pus.

Diagnosis. Appropriate treatment is only possible if a diagnosis has been made and it is generally agreed that this has become more difficult since the introduction of penicillin and the sulphonamide drugs. The acute stage of the disease is "damped down" and the condition smoulders on without producing any dramatic symptoms or signs. For this reason very careful history taking is essential and also a thorough neurological examination.

The symptoms and signs are produced in three ways:
(*a*) by the infection, local and systemic,
(*b*) by raised intracranial pressure, and
(*c*) local destruction of brain tissue and surrounding œdema

In a frontal abscess there may be a failure to improve, or a relapse, after treatment of a frontal sinusitis, with increasing headache, mental confusion, impairment of motor function in the opposite limbs and possibly speech disturbance if the dominant hemisphere is affected. Quite often an epileptic attack may be the presenting symptom.

In a temporal lobe abscess there is usually evidence of active infection in the middle ear or mastoid. The hemiplegia may be more marked than in a frontal abscess, the visual pathways are interrupted giving rise to an upper quadrant homonymous field defect or even a complete homonymous hemianopia. If the abscess is in the dominant hemisphere

sometimes it follows some surgical intervention. The organism most commonly responsible is *the streptococcus, either ærobic or anærobic.*

The infection shows itself by a widespread paralysis of cortical function, in one cerebral hemisphere, associated with a rise in intracranial pressure, high fever and sometimes rigors. There may be no frank lepto-meningitis except as a terminal event, and *lumbar puncture may show only slight changes, such as a small rise in the white cell count*



Fig 19. Osteomyelitis of frontal bone following a wound by a revolver bullet, man aged 60.

and protein content of the fluid. The patient is usually much more ill than with an intracerebral abscess and the signs are more striking. There is a flaccid hemiplegia, hemi-anæsthesia, homonymous hemianopia, and, if the dominant hemisphere is involved, aphasia. In the days before penicillin the condition was almost uniformly fatal, but the outlook is now changed.

Pus must be drained from the subdural space by burr-holes—several if necessary—while penicillin is introduced into the subdural space and the blood stream in order to combat the subdural as well as the local cranial infection and any systemic infection that there may be also. It may be necessary to give penicillin intrathecally or into the ventricles should meningitis or ependymitis be present. It is often necessary to make further burr-holes for exploration in order to drain loculated collections of pus, which are particularly liable to form on the medial and inferior aspects of the hemisphere Frequent *minor interventions of this sort are usually effective* and, as many patients are extremely ill, these are tolerated better than more major procedures.

BRAIN ABSCESS

Pathology. When an infecting organism reaches the brain the ordinary response by inflammation occurs Thus there is necrosis of tissue with liquefaction and a surrounding area of œdema. This œdema is usually more marked than with tumours. There may be a sympathetic meningitis, mild in character, with an increased protein content and cell-count in the cerebrospinal fluid, but in most cases organisms are absent.

If the patient survives the acute stage, and with modern treatment this is more likely than formerly, definite pus forms. There is a surrounding area of gliosis which proceeds to capsule formation. In abscesses of less acute development, such as are sometimes seen in

the cerebellum the capsule may be quite well developed when the case first presents, and this has a bearing on the subsequent management. At a later stage the surrounding œdema may subside and the encapsulated abscess behaves like a tumour, as a "space-occupying lesion."

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there may be an aphasia with word blindness, more marked than that seen in a frontal lesion.

In a cerebellar abscess there may be *only minimal signs of cerebellar dysfunction;* hypotonia, generalized or unilateral, ataxia, and nystagmus which is slower and coarser on looking towards the side of the abscess.

The fundi and optic disks may show papillœdema as evidence of the raised intracranial pressure. Lumbar puncture enables one to measure the cerebrospinal fluid pressure and also to analyse the fluid. There may be a slight increase of protein (chiefly globulin) and cells, and in the absence of a frank meningitis the predominant cells are usually mononuclear rather than polymorphonuclear leucocytes.

Radiography of the skull should always be done although "plain" films only rarely give much help. However they may show the site of the primary infection in the mastoid air cells, the frontal sinus or the ethmoids, or in an area of osteomyelitis. The site of a lesion may be confirmed by a shift of the shadow of the calcified pineal gland. More rarely an abscess may be revealed by a contained bubble of gas.

More information is usually obtained from the specialized radiographic investigations of angiography and pneumography of which the former is the safer and sometimes the more informative. Angiograms usually indicate the side and site of the abscess and in most cases serve to differentiate it from a tumour. This investigation causes less disturbance to the patient than encephalography or ventriculography since it does not provoke shift or other disturbance of the brain with possibly medullary compression.

Electroencephalography is of definite assistance in localizing an abscess and may actually confirm the diagnosis, as usually there is a characteristic slow, high voltage activity in the vicinity of the lesion, with phase reversal or a focus over a small area; however we have had several cases in which such a typical electroencephalogram has been shown but no abscess found on exploration, the condition presumably being a cerebral thrombophlebitis For this reason the electroencephalographic findings should be accepted with caution and only interpreted in conjunction with the whole clinical picture.

When the diagnosis has been advanced thus far it is only finally settled by an exploratory puncture. A burr-hole is made in the skull as near the site of the lesion as possible, although sometimes it may be necessary to avoid coming too close to a recent mastoid incision. A blunt (Cushing) exploring cannula is inserted and the abscess entered. If the infection is in the early necrotic stage the needle may convey the feeling of lowered resistance or absence of resistance, but if the lesion has been present some time the firm *resistance of the capsule is noted. Usually because of the associated* raised pressure, the pus escapes immediately, and should be allowed to come out slowly, without forcible aspiration, which may provoke hæmorrhage from the capsule.

Treatment. A variety of methods of treatment have been employed in the past and in most cases the subsequent introduction of new principles has produced an improvement in results over previous methods Even today, however, the mortality of cases of brain abscess is regrettably high. King (1924) introduced a method of open drainage which consisted of "uncapping" the abscess and allowing it to extrude or drain into the wound. This proved very successful in the hands of its originator but it was time-consuming and constant supervision was necessary Dandy advocated tapping (needling) of the abscess and thus tried to avoid the bad (sometimes fatal) effects of raised intracranial pressure.

Clovis Vincent performed a decompression over the abscess and also performed aspiration as frequently as necessary to tide the patient over the acute stage and enable the abscess to develop a thick capsule. The abscess was then treated as a "tumour" and excised completely.

The Edinburgh school favour continuous drainage through an indwelling catheter but in many centres it is considered that repeated aspiration is equally satisfactory, even



FIG. 20. Frontal abscess, the result of frontal sinusitis in a man of 28. Note the attachment of the abscess to the back of the frontal sinus, and the burr-hole used for aspiration

in the absence of a decompression. The general management of the case during this stage has been helped by the technique devised by Kahn (1939) of injecting "thorotrast" into the abscess at the time of the first aspiration. This makes the exact site and size of the lesion apparent in radiographs so that its progress may be watched by serial examination. Often it is possible to demonstrate enlargement of the abscess and the necessity for further aspiration before there has been any clinical evidence of deterioration in the patient's condition. At a later stage the "thorotrast" is taken up by the phagocytic cells in the capsule, which is thus outlined in the radiographs (Tutton and Shepherd, 1949). Objection has been raised to "thorotrast" on the grounds that it is radio-active and also that it produces a much thicker capsule than occurs if it is not used, although some may consider this not a disadvantage. "Diodone" (Diodrast, Pyelosil) may be used instead but as this is subsequently absorbed, the progress of the abscess cannot be followed as it can with "thorotrast."

All the above methods showed a proportion of successful results before the introduction of chemotherapy, but there is no doubt that the use of penicillin has had the effect of

increasing further the proportion of successful results. The precise role of penicillin is discussed in more detail below, but it is now routine to inject about 500,000 units of penicillin (in 1–2 ml.) at the time of the first aspiration. Although at this stage the exact organisms responsible may not be known such a dose of penicillin may have a beneficial



FIG 21 Right cerebellar abscess from chronic ear infection in a man aged 28, showing size of abscess at start of treatment Note also region of mastoid operation and burr-hole used for tapping abscess

effect by inhibiting their growth (Tutton, 1953). Thorotrast (1–2 ml.) is injected with the penicillin into the abscess cavity at the end of the first aspiration, together with 2–3 ml. of air. The patient's skull is X-rayed as soon as convenient after this to confirm the size and position of the abscess. It is sometimes found that the burr-hole is not in the best position for subsequent tapping and another can then be made in the optimum position (Figs. 20, 21). It is desirable, but not always possible, to obtain a specimen of pus for bacteriological examination before any antibiotics have been administered, as the subsequent choice of an antibiotic depends on reliable bacteriological studies. In many cases the patient has had prolonged courses of one or more of these agents and even if any organisms are present (and seen in a direct film) it may not be possible to culture them.

Subsequent aspiration of the abscess is carried out at varying intervals, the necessity usually being determined by the patient's clinical state or the radiological appearances. It is important to avoid giving the patient an excessive amount of exposure to X-rays and after the initial series it is usually only necessary to take two or at most three films at subsequent examinations.

It is sometimes necessary to carry out a second tapping of an abscess within 24 hours of the first and if so it is usually advisable to inject more "thorotrast" because that previously injected will not have been "taken up" by the capsule but lost with the pus. In most cases a dose of the appropriate antibiotic is put into the abscess cavity at each tapping. With successive aspirations the abscess usually becomes sterilized. The developing capsule may be felt with the exploring needle, but when later aspirations are performed with a sharp needle through the intact, healed scalp, it is not always possible to appreciate the resistance offered by it. This may result in the needle passing beyond the capsule and spreading infection beyond it, where another loculus may develop unsuspected and untreated, because the antibiotics do not reach it from the original abscess cavity. It is likely, however, that secondary loculi often originate from the primary source of infection quite independently of treatment.

Another risk of aspiration is hæmorrhage which is liable to occur if too forcible suction is applied, particularly in the early aspirations; it may also be caused by the sharp needle used for later aspirations. If bleeding is encountered when the needle is *in situ* it should be allowed to continue freely through the needle and in the majority of cases it will stop spontaneously. When such bleeding occurs the operator should resist any temptation to withdraw the needle and retire hastily.

As healing progresses the capsule of the abscess can be seen to contract down until only a small crenated dense shadow persists in the radiographs (Fig. 22). Opinions differ about the subsequent management but in most cases it is probably better to excise the capsule and thereby eliminate the possibility of future re-infection and perhaps also remove an unsuspected loculus. Excision is contra-indicated if the abscess is in an "eloquent" part of the brain such as the Rolandic region or the left temporal lobe in a right-handed person. Since the introduction of antibiotics some surgeons have attacked and removed the abscess in the acute stage but this probably causes more disability than waiting until the chronic encapsulated stage, as brain tissue is removed which may recover.

Treatment of Primary Focus. When the patient has been tided over the acute stage and risk to life is diminished, disease in the mastoid or paranasal sinuses or in the chest should receive appropriate treatment.

Role of Antibiotics. It has already been noted that although the newer antibiotics have been of great value in the treatment of intracranial infection they have almost certainly made diagnosis a more difficult problem and it is now exceptional to meet the classical clinical picture of a cerebral abscess with mental clouding, headache, drowsiness or coma, and clear-cut localizing signs, a slow pulse and a subnormal temperature.

There is no doubt that the widespread use of antibiotics has enabled many patients who would otherwise have succumbed to acute infective illnesses to survive and later manifest brain abscesses. The *tempo* of the infective processes is slowed and the natural defence mechanisms of the body are assisted to deal with the primary focus of infection, while in the brain itself there is restriction of liquefactive necrosis and earlier localization

of the infection. Should there be associated meningitis this should be dealt with by intrathecal injections of penicillin. It is also advisable to inject it into a lateral ventricle through a separate burr-hole at the time the abscess is tapped or when a chronic lesion is *being excised.*



FIG 22 Same case as in Fig 21. Nine months after onset only a shrivelled scar remains Note its "stalk" attached to the petrous temporal bone

The introduction of the antibiotic into the abscess cavity may sterilize it and render it inactive but we must not forget the possibility of multiple loculi which are present in nearly 50 per cent of cases, and which may remain untapped and untreated, *thus forming* foci for further trouble.

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Brain Abscess

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MENINGITIS

Different types of meningitis may come under the care of the surgeon, and may do so for a variety of reasons.

The prompt and efficient treatment of compound skull fractures is important in preventing the occurrence of meningitis. Of particular importance are those fractures in the anterior fossa of the skull, *with injuries to the paranasal air sinuses, often associated* with dural tears and cerebrospinal fluid rhinorrhea. These patients are particularly liable to develop pneumococcal meningitis and it is essential therefore in such cases to explore the anterior fossa and repair the dural defects. If the patient presents with meningitis, and it may occur quite remotely after the head injury, this should first be treated and afterwards the dural tear can be dealt with.

Although some think that systemic antibiotic treatment is adequate for the treatment of pneumococcal meningitis, we believe that it is most easily controlled by 5–7 daily intrathecal injections of 12,000-15,000 units of penicillin.

The same considerations apply in the treatment of tuberculous meningitis, and many think that a prolonged course of intrathecal streptomycin is advisable although the new antibiotics have caused modifications of traditional methods. (In the early stages of diagnosing tuberculous meningitis it is often an advantage to make burr-holes so that ventriculography can be performed to exclude other lesions such as tumour, hæmatoma, brain abscess or tuberculoma.) As a spinal block sometimes develops during the course of treatment it is helpful also to have access to the lateral ventricles through burr-holes so that the regularity of intrathecal treatment may not be interrupted.

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TUMOURS OF THE SKULL

Tumours of the skull are either primary or secondary. In the former group are the osteomas which occasionally occur in the vault of the skull and have been mistaken for the osseous reaction to a meningioma and the ivory variety which have a predilection for the bones of the orbit. Other primary tumours are the hæmangiomas, plasmacytomas and eosinophilic granulomas. Eosinophilic granulomas are met with in young subjects, usually men, produce local destruction of the bone and react well to local excision followed by irradiation (Figs. 23, 24).

Meningiomas frequently produce hyperostosis in the bone adjacent to them (Figs. 25, 26) This is a reaction in the bone due to its invasion by the tumour cells. It follows that if it is desired to replace a bone flap which has overlain a meningioma and which shows reactive changes, *it should first be boiled thoroughly to destroy tumour cells which it* may contain.

Secondary tumours in the skull may arise from malignant disease of the thyroid or from primary malignant disease elsewhere, carcinoma of the breast being the commonest but metastases from the breast are usually small and multiple whereas those from the thyroid may be single or few in number and massive in type.



FIG 23 Eosinophilic granuloma eroding skull Man aged 26.



FIG 24 Plasmacytoma in a woman of 71 The skull was eroded and the intense vascularity strongly suggested a meningioma



FIG 25. Angiogram of meningioma in region of bregma. Note vascularity of tumour, its supply by external carotid branches and also the localized thickening of the bone (hyperostosis)



FIG 26. Hyperostosis cranii produced by an angioblastic meningioma in a girl aged 16

INTRACRANIAL TUMOURS

This title is more inclusive than "tumours of the brain" and in this chapter we deal with those arising from the brain itself and from its membranes and appendages, such as the pituitary and pineal bodies and the optic nerve.

In general these lesions produce their symptoms in three ways.

(1) LOCAL IRRITATION
(2) FOCAL PARALYSIS OF FUNCTION
(3) RAISED INTRACRANIAL PRESSURE

(1) Local Irritation of the Central Nervous System

This is shown by convulsive, epileptic attacks of various types, and in the gliomata an epileptic attack quite commonly precedes all other symptoms. The attack may be generalized (grand mal) in which case it is not of any localizing value, but if the lesion is in the region of the central sulcus the attacks may be mainly motor convulsions without loss of consciousness ("Jacksonian") or there may be subjective sensory phenomena, such as paræsthesia, sometimes called sensory Jacksonian attacks. If the seizure originates in the region of Broca's area there may be a temporary disturbance of speech function during or after an attack. Occasionally after a major convulsion there is a temporary paralysis of an extremity (Tod's paralysis). As this may recover quite rapidly it is important to observe it, if at all possible, as it will give valuable help in localizing the lesion.

Lesions in the medial part of the temporal lobe may cause uncinate attacks in which there are olfactory or gustatory hallucinations, associated with a sensation of unreality, confusion and automatic behaviour. Temporal lesions may also be associated with vivid, formed, visual hallucinations or auditory hallucinations.

(2) Focal Paralysis of Function

The temporary weakness of a Tod's paralysis may in time become permanent and progressive weakness of an extremity develop. Tumours in the upper and more medial part of the motor strip produce more effect in the lower limb while those in the lower part of the frontal lobe affect predominantly the face and upper limb. In parietal lesions impairment of function and apparent paralysis may be due to sensory deficit rather than motor weakness.

In lesions around the base of the skull there may be interference with the various cranial nerves proudcing anosmia, visual field defects or ocular palsies.

Where there is pressure on the chiasm, as by a pituitary tumour, there is a bitemporal hemianopia, the early field loss starting in the upper quadrants and progressing until the temporal half of the field is completely involved. If the pressure is unrelieved the lower nasal fields become involved and later complete blindness may result. The changes may not be symmetrical in the two eyes and this is particularly so in compression of the chiasm from lesions such as aneurysms or suprasellar meningiomas, where also the pattern of progress may differ. When the optic tract or radiation is involved there is a homonymous hemianopia. In an early temporal lobe lesion only the lower fibres of the radiation may be involved where they pass around the temporal horn of the lateral

ventricle (Meyer's loop), in which case only the upper quadrant will be affected. On the other hand, a parietal lobe lesion may affect only the upper fibres of the radiation causing a lower quadrant homonymous field defect. The more advanced lesions of the optic radiation cause complete homonymous hemianopia.

The associated neurological deficit may be of value in deciding the more precise situation of the lesions which produce these different visual field defects and serial observations detect early changes which enable progress to be assessed and localization to be more exact.

Involvement of the trigeminal nerve or its ganglion produces sensory loss in the corresponding half of the face and forehead. In lesions below the tentorium other cranial nerves may be affected, from the seventh to the twelfth inclusive.

Infiltrating lesions developing in the frontal lobe and tumours in the third ventricle may produce marked alterations in personality, the presence of which may only be revealed by the independent testimony of the patient's relatives. The patient may have become careless and dirty in habits or there may be impairment of memory or powers of concentration. There may be unnatural cheerfulness, with a lack of insight into the serious nature of his condition, and in some cases an unusual facetiousness ("Witzelsucht"). Occasionally these patients are incontinent when fully conscious and remain quite unashamed and undisturbed by the discomfort of a soiled bed.

Some blunting of mental faculties may be caused by the general increase in intracranial pressure and so the exact sequence of events in the history may be of great importance.

(3) Raised Intracranial Pressure

This is produced either by the progressive increase in size of the tumour or by obstruction of the cerebrospinal fluid pathways at one or other of the "narrows" such as the foramen of Munro, third ventricle, iter, or fourth ventricle. Raised pressure, however produced, causes headache, drowsiness, nausea, vomiting and slowing of the pulse. These pressure phenomena are frequently most striking in the early morning when the patient wakes and depend on the rise in intracranial pressure which occurs overnight during sleep. Vomiting without headache is seen in lesions in or near the fourth ventricle.

The clinical picture produced by a tumour depends both on its effect on the cerebrospinal fluid circulation (the third circulation, Cushing) and on its effect on the blood supply to the adjacent brain; thus a small tumour in the posterior fossa by obstructing the flow of cerebrospinal fluid may produce high intracranial pressure in a comparatively short time while a large tumour may be present in the frontal region with little or no effect on intracranial pressure. This is particularly so in older people where the shrunken brain allows more room for accommodating the growth of a tumour, the progress of which may be quite silent and asymptomatic. The effect on blood supply depends again on the rate of growth of the tumour; if this is slow a collateral circulation may develop, e.g. a large meningioma may be present in the Sylvian fissure without hemiplegia whereas a smaller rapidly growing tumour in the same region may produce profound hemiplegia within a short time. The large meningioma has grown so slowly that local cerebral anæmia has not occurred because a collateral circulation has been established coincidently with its development.

SPECIAL INVESTIGATIONS

There are several specialized techniques used in the investigation of intracranial tumours and these are described before giving detailed consideration to the different lesions and their signs, symptoms, and treatment.

In all cases it is essential to take a careful clinical history. Owing to the impairment of consciousness or cerebration in many patients, it is usually necessary to interview their relatives in order to obtain a satisfactory history. In this way valuable information may be obtained regarding alterations in personality, mental retardation and disturbances of speech or memory. Even when a patient is apparently quite alert and co-operative it is sometimes striking how much at variance is the history given by him with that by close relatives The time relationships of the different symptoms may give an indication of the speed of growth and make a pathological diagnosis easier.

A careful general examination must obviously be carried out in addition to the detailed examination of the central nervous system. This enables the patient's fitness for operation to be assessed and may also reveal an undisclosed primary tumour when an intracranial metastasis is producing the clinical picture. The commonest site for such a primary lesion is the bronchus, but carcinoma of the prostate, breast, thyroid, stomach, kidney, adrenal, testis, colon and skin (melanotic carcinoma) may also spread in this way.

The examination of the olfactory nerves, although often omitted, should not be overlooked. There is sometimes loss of the sense of smell after fractures involving the anterior fossa of the skull, particularly when there has been an associated dural tear with cerebrospinal fluid rhinorrhea. There is also loss of the sense of smell in meningiomas arising from the "olfactory groove" and rarely also in cases of frontal glioma.

The careful testing and recording of the visual acuity is important as it not only gives a quantitative record of visual failure but also enables the observer to assess accurately any subsequent improvement or deterioration. The examination of the visual fields should never be neglected. In addition to the examination of the peripheral fields on the perimeter the central vision should be carefully charted on the Bjerrum screen at a distance of 2 metres On the whole it is the screen examination which yields most help. As a rough test in the out-patient department or at the bedside the method of confrontation can be used, but although this may reveal gross defects it is possible to miss small field defects which may be of great importance in localization. "A negative finding with a rough test only shows the absence of a gross hemianopia and should not be allowed to stand in the way of a more adequate investigation" (Traquair, 1948).

Increased intracranial pressure shows its presence by its effect on the optic disks and ophthalmoscopic examination may reveal papillœdema of varying degree, although it is by no means constant and may be entirely absent

Significant findings in the remainder of the neurological examination are mentioned with the different types of tumour.

The head should be carefully examined, particularly after shaving, when abnormal vascularity, localized bulging and thinning may be seen or felt. The circumference of the head should be measured This will not only give information in cases of hydrocephalus and infantile subdural hæmatoma, but may also reveal abnormal enlargement in children, where separation of the sutures may permit quite a large tumour to be accommodated before any symptoms of raised intracranial pressure develop. In these cases percussion

also is of value and a characteristic high-pitched note (cracked-pot note, or Macewen's sign) may be obtained, especially on percussing near to the sutures.

Auscultation of the head will reveal a bruit in some cases of tumour, aneurysm or angiomatous malformation (Mackenzie, 1955); in conditions such as carotico-cavernous fistula the maximum bruit may be heard over the globe of the eye. Lumbar puncture will enable the pressure of the cerebrospinal fluid to be measured and examination of the fluid may show increased protein, increased cells or even the presence of tumour cells, but its risk in cases of high intracranial pressure as shown by highly choked optic disks must not be forgotten.

X-ray Investigations

X-ray examination of the skull is indispensable, and in certain instances may not only confirm the presence of a tumour but also indicate its site and its nature. Some abnormality will be demonstrated in at least 60 per cent of cases. The general signs caused by raised intracranial pressure are, separation of the sutures (particularly in the young), "beaten brass" appearance and decalcification of the dorsum sellæ, leading in some cases to its complete disappearance (Figs. 25, 27, 32, 34) Other possible findings in a "straight" film include localized thickening or erosion of bone, increased vascularity, areas of abnormal calcification, displacement of the calcified pineal gland, enlargement of the sella tursica (Fig. 33), of the optic foramen or of the porus acusticus internus In view of the possibility of the intracranial lesion being metastatic, radiography of the chest should never be omitted

Pneumography

Further help can be obtained by the use of contrast media in radiography, particularly air which can be injected by lumbar puncture (encephalography) or directly into the ventricles through burr-holes made in the parietal or frontal region (ventriculography) (Fig. 28 (*a*), (*b*)) The disadvantage of ventriculography is that it may cause considerable disturbance to the patient by an acute rise in intracranial pressure. For this reason it is better to carry out this investigation only when everything is prepared for immediate craniotomy should this become necessary. In general, encephalography should not be used where there is markedly raised intracranial pressure as shown by the presence of papillœdema, but the Swedish school has shown its value, if carefully done with small quantities of air, even in lesions of the posterior fossa

Occasionally when good visualization of the third ventricle, aqueduct and fourth ventricle is of vital importance a positive contrast medium ("Myodil" 1–2 ml.) is introduced into the lateral ventricle through a burr-hole and brought through under direct screen control, to the posterior parts of the ventricular system. It has been of particular value in certain cases, where an anomaly such as a very cellular mastoid makes air pictures unsatisfactory

Angiography

Much less disturbing is the other contrast investigation (angiography) in which a radio-opaque substance is injected into the carotid or vertebral artery and serial exposures are made to show the filling of the intracranial arteries, capillaries, veins and sinuses. Formerly it was necessary to expose the artery but a technique has now been developed of puncturing the artery percutaneously, which makes the investigation more

easily performed and more acceptable to the patient; it is even possible to apply it to out-patients When cerebral angiography was first introduced by Moniz in 1927 it was thought that it would be principally applicable to the diagnosis of vascular lesions such as



FIG 27 Intense vascularity of skull in large parietal meningioma Note destruction of sella tursica from long-standing raised intracranial pressure

aneurysms, but the advances made in its development in Scandinavia, and later in this country and the United States have shown its great value in the diagnosis of space-occupying lesions, such as tumours, abscesses and hæmatomas (Figs. 6, 25, 29).

It should be emphasized that angiography is complementary to pneumography and has not displaced it as a method of investigation, there are particular indications for each, and each has its own particular value. When a lesion has been lateralized to one hemisphere angiography is the method of choice but in suspected deep-seated tumours, or those in the mid-line or posterior fossa ventriculography is preferable.

Diagnostic Burr-hole

In the detailed investigation of an intracranial lesion it is not easy to draw a line between investigation and treatment and it is sometimes necessary to make a diagnostic burr-hole and to carry out exploratory needling. The former is especially useful in



(a) (b)

Fig 28 (a) *and* (b) Ventriculograms in large frontal glioma. Note the shift of the ventricles in the antero-posterior view, whilst in the lateral projection there is also backward displacement of the affected ventricle

confirming the presence of a hæmatoma (extradural or subdural) but may also help in the localization of a tumour; the latter may reveal the existence of an intracerebral hæmatoma, abscess, cyst or tumour. When a tumour is present it is usually possible to recover from the eye of the cannula a fragment of tissue which can be spread on a slide and after suitable staining will enable the histological diagnosis to be confirmed. In this way a major intervention may be avoided in unfavourable cases, but care is necessary in the interpretation of results, as the astrocyte reaction at the edge of an abscess or meningioma may be wrongly reported as a glioma (Russell, Krayenbuhl, and Cairns, 1937; Russell, 1951).

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TYPES OF TUMOUR

The Gliomas. These form nearly one half of all intracranial tumours. They arise from the neuro-ectodermal cells which form the supporting tissue of the central nervous system, although neuronal elements are also seen in some of them. Of the various tumours in this group approximately 80 per cent are of three main types, viz. (1) glioblastoma multiforme, (2) astrocytoma and (3) medulloblastoma.

(1) The **glioblastoma multiforme** occurs mainly in the fourth to the sixth decades. It is a rapidly growing cellular tumour and in its growth tends to outstrip its blood supply so that there are often necrotic or pseudocystic areas in its substance. It usually infiltrates the surrounding brain beyond the area where there is obvious macroscopic tumour and this makes it difficult to effect a radical excision.

The length of history is rarely more than a year and most commonly is much less. The onset may be heralded by an epileptic attack or there may be slowly developing changes in personality and temperament, or a hemiplegia. In many of these cases the diagnosis may be confirmed by angiography (Fig 29).

Operation has little to offer sufferers from these tumours and in some clinics it is customary merely to confirm the diagnosis by a needle-biopsy (*vide supra*) through a suitably placed burr-hole, but if the tumour is in the frontal lobe, the right temporal or the occipital lobe, radical excision may be attempted and subsequently a course of radiotherapy given. A wide excision of the tumour provides the patient with an "internal decompression" and may postpone the almost inevitably fatal outcome Not uncommonly these tumours occur in patients over 60 years of age and as they may be slow to produce a rise in intracranial pressure they may be overlooked An epileptic attack or slowly developing hemiplegia in such a patient should not be dismissed as a vascular accident or hypertensive encephalopathy until after a thorough investigation. This should include examination of the cerebrospinal fluid, particularly its pressure and protein content, electroencephalography and careful radiography with, if necessary, angiography or

air studies. Occasionally a patient's symptoms are found to be caused by a chronic subdural hæmatoma in spite of the complete absence of any history of injury (see p 19)

(2) **Astrocytomas.** These form about one-quarter of all gliomas and two main types are described. In adults from 30-50 the tumour occurs as a slowly growing diffuse



Fig. 29. Angiogram of malignant glioma in a man of 39 Note vascular displacement, irregular filling of abnormal vessels and rapid "shunting" into venous circulation

infiltrating lesion in the cerebral hemispheres, occasionally with cyst formation and, less commonly, areas of calcification. It tends to infiltrate among the normal fibre and cell structures of the brain without interference with function, so that it may become quite extensive before causing symptoms. Sometimes there is a history of an isolated epileptic attack or series of attacks occurring several years before any other symptoms develop. There is evidence that anaplastic changes may occur in a slowly growing astrocytoma causing it to develop into the more malignant, rapidly growing, invasive, glioblastoma multiforme.

In children and young adults the astrocytoma occurs mainly in the cerebellum and is usually well demarcated and may be associated with cyst formation. It often occupies the vermis although, when large, it may extend laterally into one or both cerebellar hemispheres. When these cases were first treated only the cyst was tapped but symptoms recurred after a variable period and after re-exploring it was found that the essential part of the operation was to remove the mass of solid tumour (mural nodule) in the wall of the cyst. Because of their slow growth they may reach surprising dimensions before the

patient presents for treatment with the consequence that the skull often shows marked evidence of raised intracranial pressure with separation of the sutures and erosion of the dorsum sellæ. Air studies show internal hydrocephalus which may be of severe degree (Fig. 30).*

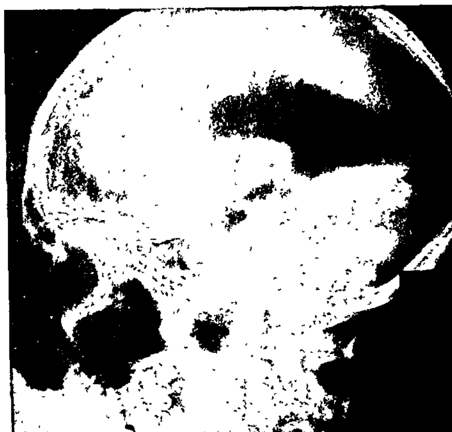


FIG 30 Ventriculograms showing marked ventricular dilatation with kinking and dilatation of the aqueduct Cerebellar glioma in a boy of 18.

(3) **Medulloblastoma.** This highly malignant tumour occurs in the cerebellum of children and is usually in the midline in the region of the roof of the fourth ventricle. It forms a soft, greyish, friable mass and tends to spread through the subarachnoid space, so that greyish plaques of tumour may appear over the surface of the cerebellar hemispheres or the spinal cord, remote from the main mass. As a rule patients with medulloblastoma are younger than those with astrocytoma although this does not always help in the diagnosis of an individual case. They present a similar clinical picture of raised intracranial pressure and cerebellar dysfunction. Microscopically the tumour consists of small, closely packed cells with densely staining nuclei which may be grouped to form rosettes. Mitoses are numerous and occasional multinucleate cells are seen. As might be expected these tumours are highly radiosensitive and, after pathological confirmation together with an appropriate decompression, a thorough course of irradiation

* The prognosis in cerebellar cystic astrocytoma is good and one of our patients upon whom we operated 22 years ago, afterwards served throughout the war in the W.R.N.S and has married and become the mother of several children.

offers the best chance of survival, but even under the best possible conditions the prognosis is poor.

Less commonly seen gliomas are ependymoma, oligodendroglioma and spongioblastoma polare.

The **ependymal tumours** arise in some part of the ventricular system, most commonly in the fourth ventricle in children and young adults. When they occur in the lateral ventricles they are frequently calcified (Bailey, 1948) They are slowly growing and relatively benign, although they sometimes have attachments to the floor of the fourth ventricle which makes a complete excision difficult or impossible. They are firm in consistence, avascular and easy to handle at operation, but from their position it may not always be wise to attempt removal. A less differentiated and more malignant type of this tumour, the ependymoblastoma, is described but is very uncommonly seen.

Oligodendrogliomas form about 4 per cent of all gliomas and occur mainly in the cerebral hemisphere of young adults and children. They are slowly growing and fairly well circumscribed and are most characteristically the tumours which show calcification, so that they may be identified in a plain radiograph. They can usually be excised completely and show little tendency to recur. Exceptionally they show more malignant, invasive tendencies and may spread through the subarachnoid pathways in a manner reminiscent of the medulloblastomas (Beck and Russell, 1942).

A slowly growing infiltrating type of glioma occurs in the optic chiasm, the corpus callosum and pons (diffuse hypertrophy of the pons). Although some authorities have called this a **polar spongioblastoma** others consider it a variety of astrocytoma known as "piloid," which on section shows bundles of slender cells with neuroglial fibrils.

Meningiomas.* These tumours constitute about 15 per cent of all intracranial tumours and they arise from arachnoidal cells (meningocytes) commonly in relation to the large venous sinuses, although they do develop in other places. The common sites are parasagittal (and falx), over the convexity of the cerebral hemispheres, the sphenoidal ridge, olfactory groove and the tuberculum sellæ. Less frequently they occur in the lateral ventricle and in the posterior fossa. There is evidence that in some cases trauma has played a part in determining their development, particularly in those which arise in the region of the bregma.

The tumours are usually closely attached to the dura (Fig. 31) but displace the adjacent brain and produce a clear line of separation although occasionally they are irregularly nodular and appear to be infiltrating. They are usually roughly spherical in shape and on account of their slow growth and the adaptability of the brain may reach quite large proportions, particularly in the frontal region, before they present for treatment. A meningioma may invade the adjacent bone and produce overgrowth and thickening (hyperostosis) which may be demonstrable in a radiograph and give a clue to the diagnosis. Such bony changes may be seen on the sphenoidal ridge, tuberculum sellæ or olfactory groove and also over the vault of the skull where the thickening may extend internally (endostosis), externally (extostosis) or both (Figs 25, 26). More rarely areas of calcification may be seen in these tumours (Fig 32) or they may be outlined by a ring of calcification but they are most commonly demonstrated by angiography (Fig. 25).

* These [illegible]
by Harvey [illegible]
called mas [illegible]
origin of the neoplasm.



FIG. 31. Meningioma weighing 96.2 grammes removed from left parasagittal region of a woman aged 32.



FIG. 32. Calcified meningioma arising from region of anterior clinoid process in a man aged 46.

They are essentially benign, but all observers who have had the opportunity to follow their cases over a number of years have noted the significant number which have recurred. Although the tumours are circumscribed they frequently have extensive vascular connexions, as much from the meninges as from the cerebral vessels. This vascularity may make excision quite a formidable procedure and in some cases operation in stages may be necessary. The tumour may actually infiltrate the sagittal sinus a portion of which may need to be excised but if, as is most likely, the sinus is already obstructed no ill effects are likely to follow (Jaeger, 1942).

Cerebellar Hæmangioblastoma. This tumour arises in the cerebellum or medulla oblongata most commonly in young adults aged from 30–40 years. It arises from vascular primordia which are closely connected with the pia and consequently is nearly always near the surface of the cerebellum. Recent work suggests that this tumour is closely related to the angioblastic meningioma. It is sometimes solid, but more commonly there are numerous small cystic and hæmorrhagic spaces, or one large cyst containing yellow fluid with the actual tumour forming a small, yellowish or red mural nodule. Occasionally there is an associated angiomatosis retinæ (von Hippel's disease) and cystic changes in the pancreas and kidneys with benign tumours of the kidneys and suprarenal glands (Lindau's syndrome), the changes sometimes showing a familial incidence. There may be similar vascular tumours in the spinal cord.

The symptoms produced are those of any other cerebellar tumour but the exact diagnosis may be made if an associated retinal hæmangioma is discovered. The treatment consists of removal of the tumour, and the essential part of the operation is excision of the mural nodule which, as it is commonly situated posteriorly and on the surface of the cerebellum, is usually easily performed. Careful follow-up over a long period has shown that quite a number recur but it is suggested that recurrences of these tumours are usually not due to incomplete excision but are actually second tumours arising *de novo*. Their occurrence in a different site would lend support to this hypothesis.

Acoustic Neurinoma. This tumour develops from the perineurium of the acoustic nerve in the region of the internal auditory meatus. As the tumour enlarges it occupies the cerebello-pontine angle and indents the adjacent cerebellum and brain stem. The tumour usually has a well developed capsule, is occasionally cystic and quite frequently has an arachnoid cyst overlying it. It produces symptoms by interference with the eighth nerve (tinnitus and progressive loss of hearing), the facial and trigeminal nerves and by pressure on the cerebellum. Tumours on the lower cranial nerves are sometimes seen as part of a generalized neurofibromatosis, when they may be bilateral or multiple. Histologically the neurofibroma is different from the solitary acoustic neurinoma (Schwannoma) but from the clinical point of view behaves similarly. The tumours tend to remain undiagnosed in their early stages and their intimate relation to the brain stem and adjacent cranial nerves often makes their removal difficult. It may be impossible to avoid sacrificing the facial nerve but this sacrifice is justifiable if a complete removal can be performed, as operations for recurrences carry a significantly higher mortality. If the facial nerve is permanently damaged some function may be restored to the facial muscles by a nerve anastomosis in which the proximal part of the spinal accessory or the hypoglossal nerve is joined to the distal part of the divided facial. We have found it preferable to use the former, as paralysis of the sternomastoid and trapezius muscles seems less of a disability than a paralysed and wasted half of the tongue.



Fig 31. Meningioma weighing 96 2 grammes removed from left parasagittal region of a woman aged 32



Fig 32 Calcified meningioma arising from region of anterior clinoid process in a man aged 46

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In addition to the characteristic clinical findings, with the auditory nerve tumours it is usual to find a considerably raised protein content of the cerebrospinal fluid. This is so constant that one is reluctant to diagnose an "angle" tumour if the protein level is less than 100 mgm. per 100 ml. In an advanced case there may be raised intracranial pressure, with papillœdema, due to internal hydrocephalus produced by obstruction in the posterior fossa and in such cases it might be unwise to perform lumbar puncture to ascertain the pressure and the protein content unless it is done in the theatre immediately before operation.

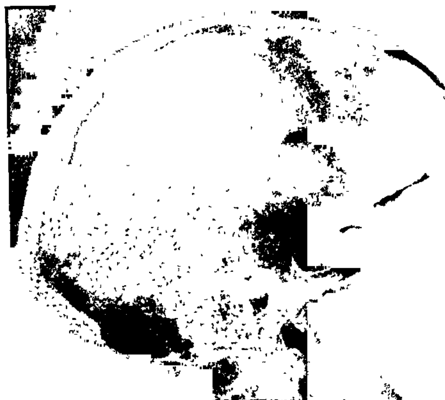


FIG 33 Chromophobe adenoma of pituitary in a man of 38. Note enlargement of sella tursica and displacement of air-filled third ventricle and basal cisterns.

Tumours of the Pituitary

These constitute about 10–15 per cent of primary intracranial tumours and are adenomas of the anterior lobe. Tumours arising from the posterior lobe are virtually unknown.

Chromophobe Adenoma. This is the commonest pituitary tumour and is so called because its cells generally contain no granules which take up the characteristic stains (Fig. 33). They are about three times as common as the eosinophil (acidophil) adenomas which are associated with gigantism in the growing subject and acromegaly in the adult and which do not grow to the same size as the former and much less commonly produce signs of pressure on the optic chiasm.

The chromophobe adenoma typically extends upwards, destroying the sella and produces pressure on the optic chiasm, the visual fields showing a bitemporal hemianopia. In addition there are endocrine disturbances with loss of sexual activity and interference

with water metabolism, shown by polyuria and polydipsia. The patient may show a characteristic appearance with a smooth skin, fine soft hair on the head and an absence of body hair generally. Male patients require to shave only infrequently and are often impotent; women may have irregular, scanty menses or amenorrhea.

Occasionally these tumours grow beyond the sella and extend into the temporal or frontal lobes, producing interference with the various ocular nerves or obstruction of the third ventricle.

In young persons with posterior fossa tumours and internal hydrocephalus a spurious picture of pituitary dysfunction may be produced, with obesity and other endocrine disturbances, and chiasmal compression, solely due to secondary pressure effects from the dilated third ventricle.

Treatment of patients with pituitary adenomas is principally aimed at the preservation of vision and in most chromophobe tumours operation is necessary to relieve the pressure on the optic chiasm. This also allows histological confirmation of the nature of the tumour which enables a decision to be made regarding subsequent radiotherapy. On the whole it is inadvisable to irradiate these tumours without preliminary operation, as tumours insensitive to X-rays or even an aneurysm may produce a chiasmal syndrome and mimic a true pituitary tumour.

Histologically pituitary tumours sometimes show a mixture of cell types and the clinical appearance may also be atypical, e.g. a patient with a syndrome suggesting a chromophobe adenoma may show a slight but definite acromegalic appearance.

Acidophil (Eosinophilic) Adenoma. This seldom grows to the size reached by the previous tumour and thus only exceptionally produces pressure on the chiasm, but in some cases the tumour becomes enormous and invasive and rapidly destroys the life of the patient. The sella tursica is often ballooned downwards while its upper part, including the diaphragma sellæ remains intact. It is suggested that stretching of this latter structure may be responsible for the headaches which are often severe in the early stages of cases of pituitary tumour. If this tumour develops before puberty it produces gigantism, but in the adult it causes acromegaly. As the usual indication for operations is to relieve chiasmal compression it is unusual for these patients to require operation. Radiotherapy is often of value, although the bony changes are permanent and not affected by treatment. Beneficial effects have been derived from treatment with œstrogens, which are said to inhibit the growth of the acidophil cells in the pituitary.

Basophil Adenoma. This is a much less common lesion and on the whole it is not of surgical importance. It is probably responsible for Cushing's syndrome which is characterized by obesity, hypertension, plethora and cutaneous striæ. There is not usually any evidence of chiasmal compression. A similar clinical syndrome is produced in some cases of neoplasm of the cortex of the suprarenal or of the thymus. Estimation of the 17-ketosteroids excreted in the urine may help in the differential diagnosis, the concentration being greatly increased in adrenal tumours.

Tumours of the Hypophyseal Duct. (Syn: suprasellar cyst, cranio-pharyngioma, adamantinoma, Rathke pouch cyst.)

These tumours arise from remnants of the craniopharyngeal pouch and although most commonly they develop above the sella they may occasionally form intrasellar tumours. They occur usually in young subjects and the symptoms produced depend on their exact site of origin, but pressure on the chiasm is usually present and visual defects



(a)



(b)

FIG. 34 (a), (b) A large suprasellar cyst has been aspirated and air injected Note also the separation of the coronal suture, the "beaten brass" appearance, the destruction of the sella tursica and the calcification above it.

develop. The presenting symptom is often progressive failure of vision. Interference with the pituitary gland causes delay in growth and physical development, with sexual immaturity, or alteration of sex function if puberty has already been reached. There may in addition be episodes of diabetes insipidus, alterations of sleep rhythm, emotional outbursts and obesity. The tumour is usually solid in the early stages but later a cystic component develops and this may form its main mass (Fig. 34 (*a*), (*b*)) The cyst fluid which contains cholesterol crystals is usually yellow, or turbid and brown resembling engine oil, and there is often some degree of calcification in this lesion, varying from a faint streak in the wall of the cyst to a very dense mass in the solid part of the tumour. It spreads upwards into the third ventricle and under the frontal lobes. Eventually there is obstruction of the foramen of Monro with development of internal hydrocephalus and evidence of raised intracranial pressure.

Operation is usually indicated to relieve chiasmal compression. Temporary relief from symptoms can sometimes be obtained by aspiration of the cyst through a burr-hole and an injection of air at the same time will outline the cyst and show its true dimensions. Radical removal is not usually an easy matter on account of the close relationship that develops with the walls of the third ventricle (hypothalamus) and even under the most favourable circumstances there is a substantial mortality. The use of A.C.T.H. and cortisone have helped to minimize the hypopituitary and adrenal crises which may develop after these operations.*

Pituitary Abscess. These cases are of unusual and of obscure pathology. We have encountered one in a seaman aged 38 (Rogers, 1949) from whose pituitary region we removed a large quantity of creamy sterile pus consisting of polymorphonuclear leucocytes and debris. Other cases have been reported by Frazier (1930) and Svein and Love (1942).

Pinealoma. Tumours developing from the pineal gland are rare. Histologically they are characteristic and resemble the seminoma of the testis. Evidence has been produced to show that they may be atypical teratomas and in males there is sometimes pubertas præcox. By their growth they produce pressure on the quadrigeminal plate and obstruction of the aqueduct. The typical clinical finding is impairment of upward conjugate movement of the eyes but there may also be pupillary changes with dilated inactive pupils but preservation of the accommodation reaction. Downward extension may produce cerebellar disturbances due to interference with the superior cerebellar peduncles and pressure on the medial geniculate bodies may cause deafness. Ventriculography shows an internal hydrocephalus with a filling defect at the posterior end of the third ventricle.

These tumours can be approached by splitting the splenium of the corpus callosum but a complete removal is often impracticable. It may be better simply to obtain a biopsy specimen to confirm the histology, perform a ventriculocisternostomy (Torkildsen, 1947) to relieve the obstruction of the cerebrospinal fluid pathways and then irradiate the tumour.

Intraventricular Tumours

As described above meningiomas and ependymomas sometimes occupy the ventricles but two other tumours occur very typically in this situation.

* These tumours may occur in adults and we have suggested (*J. Hist. Med. & Allied Sciences*, 1949, 4, 468) that Milton's blindness may have been caused by one of them

Colloid Cyst of Third Ventricle. This is a rare tumour which develops in the anterior end of the third ventricle from the paraphysis, a vestigial structure in the dorsal part of the lamina terminalis. As the tumour enlarges it blocks the foramina of Monro, but in the early stages this obstruction may be intermittent and the patient suffer from periodic acute attacks of raised intracranial pressure ("hydrocephalic attacks"). These may start quite suddenly and be relieved by an alteration in position of the head. There is usually a characteristic ventriculographic picture with sometimes complete absence of filling of the third ventricle. The tumour may be approached through the frontal lobe and across the dilated lateral ventricle. Dandy found a posterior approach through the third ventricle to be equally satisfactory. The tumour has few attachments, is easily removed and shows no tendency to recur.

Papilloma of the Choroid Plexus. This may be found in the lateral, third, or fourth ventricle and may be associated with excessive production of cerebrospinal fluid and internal hydrocephalus. There is usually a raised concentration of protein in the cerebrospinal fluid and tumour cells may sometimes be demonstrated in it. These scattered tumour cells may develop elsewhere in the meninges as seedlings, but the tumour itself is histologically benign. When in the lateral or fourth ventricle it is usually amenable to removal.

Metastatic Tumours. These are frequent, the primary sites being most commonly lung and bronchus, breast, stomach and bowel. Less often they originate in prostate, kidney or skin (melanotic carcinoma).

The tumours may be solitary and are found in both the cerebellum and the cerebral hemispheres. They tend to occur at the junction of grey and white matter and are usually circumscribed and easily removable. In some cases the secondary tumour presents before there is any hint of the presence of the primary growth. Sometimes, even at post-mortem, a very careful search may be required to find a small primary lung tumour, even when the secondary deposit has been readily identified. Operation may be justifiable in order to relieve pressure or preserve vision, and in rare cases it may be possible to attack the primary lesion afterwards.

Granulomas. Tuberculomas have never been common in this country, although numbers of cases have been described in Scotland and Southern Ireland. They are much more frequent abroad, in the Iberian peninsula and in South America. Occasionally they are calcified but quite commonly they are found during an exploration for a suspected tumour, usually one in the posterior fossa. Until recently any attempt at their removal was likely to be followed by fatal meningitis, but since the introduction of streptomycin this risk has been greatly reduced and the outlook has correspondingly improved. However, since a number occur in sites, such as the brain stem, from which removal is not practicable, and since many patients have tuberculous lesions elsewhere, the condition remains a very serious one.

Gummatous tumours are very rare and are thus not usually diagnosed before operation. Signs of an intracranial tumour in a patient with a positive Wassermann reaction are more likely to be due to a primary neoplasm, than a gumma. The lesions commonly arise in the meninges but later spread into the brain and may be difficult to separate cleanly from it. They are resistant to antiluetic treatment.

Infection of the central nervous system may be caused by a yeast-like organism, torula histolytica, and although most commonly it causes a diffuse chronic meningo-

encephalitis it may also give rise to a chronic granulomatous mass called a toruloma. The tumour may occur in either the cerebrum or the cerebellum and examples of their successful removal are recorded.

Parasites

Hydatid disease is endemic in Wales and, particularly in younger people, has to be considered in the differential diagnosis of intracranial and spinal tumours. Ancillary investigations include the Casoni and complement fixation tests and the eosinophil count and these should help in reaching a diagnosis. The cysts develop slowly and may reach quite a large size, producing enlargement of the head, *localized or generalized*, before announcing their presence. With careful operative technique it is possible to excise them completely without risk of recurrence (Arana-Iniquez and San Julian, 1955) (Langmaid and Rogers, 1940) (Rogers, 1955).

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[illegible] 23, 656.

New Orleans.

CEREBRAL ANGIOMATA

Although these were originally thought to be tumours the majority are arterio-venous anomalies, almost certainly congenital in origin. Very few have true tumour tissue in them (hæmangioblastomas). They were thought to be uncommon lesions until the increased use of angiography showed them to be relatively common. Various types have been described, with either arterial, capillary or venous elements predominating, but they are all fundamentally the same, the essential part of the lesion being the multitude and tortuosity of the vessels and the arterio-venous shunt between them. All other changes are secondary to this. The tendency is for the blood flow to be diverted into this easier pathway instead of passing through the normal cerebral vessels so that part of the brain may be deprived of its normal blood supply. This is shown by the fact that when such a lesion has been excised there is an improvement in the filling of the normal cerebral vessels (Fig. 11, p. 26).

Symptoms

There is often a long-standing history of headache, which may be unilateral and regarded as migraine while in most cases fits, which are usually focal but may be generalized, are a prominent part of the clinical picture. The tendency is for the lesion to be progressive and successive fits may leave the patient with a weakness of a limb, progressing to a monoplegia or hemiplegia. There is, however, great variability in the frequency

and severity of the attacks and there may be quite long periods of freedom. Associated with this march of events there may be some degree of mental disturbance, sometimes accentuated by the frequency of the fits or the progress of a paralysis.

Hæmorrhage is a common complication and in many instances may be the presenting symptom. It is usually less severe and devastating than that into the subarachnoid space from a ruptured aneurysm and there may be many recurrences without a fatal issue. In some cases a spontaneous cure has followed although in other instances each hæmorrhage seems to be associated with some progress of the lesion. If there is bleeding into the hemisphere there may be loss of consciousness, hemiplegia, hemianopia or aphasia depending on the particular region of the brain involved. Clinically such cases may resemble subarachnoid hæmorrhage from an aneurysm or acute œdema from a rapidly growing glioma, but differentiation may be made and the diagnosis established by angiography.

Diagnosis

This may sometimes be suspected from the history. In a proportion of cases, which has varied in different series, an intracranial bruit may be heard, but even this is not absolutely pathognomic of an angioma (Mackenzie, 1955). Radiographs of the skull sometimes show distinctive calcification in the form of parallel strips ("tram lines") in these lesions but in most cases angiography is necessary to establish diagnosis. In the more posteriorly situated lesions vertebral injections should also be done as there may be an important contribution from the posterior cerebral artery. Intelligent use of this procedure is necessary as the lesion may only be demonstrable during one short phase of the intracranial circulation and thus may be missed if only a small number of routine exposures are made. Curtis (1949) described one that filled at a very late stage and was only demonstrated by serial angiography.

Treatment

It is only in recent years that these cases have been considered as suitable for surgical treatment. Previously when exposed at operation they were either left strictly alone or cautious efforts were made to ligate some of the vessels or to coagulate them with the endothermy. Radiotherapy was sometimes given subsequently and although it was usually ineffective there is evidence to suggest that a small number of patients derived benefit. Carotid ligation was also practised but never produced more than minor improvement, for it could have little effect on the main lesion. Angiography has enabled the lesion to be carefully studied before operation and the Swedish school of neurosurgeons led the way to the carrying out of complete excision (Olivecrona *et al.*, 1948). Excision is not possible in all cases but improvements in technique and experience will enable more to be radically treated. Extirpation not only removes the risk of subsequent hæmorrhage but may also reduce or abolish epileptic attacks. This gain to the patient must sometimes be balanced against an increase in the physical disability but many patients think this a small price to pay for the freedom from fits and protection from the danger of hæmorrhage. In general these lesions should be either excised completely or left alone.

As with the saccular aneurysms the use of arterial hypotension and hypothermia has made them less formidable, but the surgeon "must occasionally have the courage and judgment to withdraw after actual exposure of the lesion" (Pilcher, 1946).

Spontaneous Intracerebral Hæmatoma

Young adults sometimes develop a progressive hemiplegia, possibly with hemianopia and aphasia, but without necessarily any loss of consciousness. This has been found to be due to an intracerebral collection of blood which may be frontal or parietal in situation. Angiography often does not show an aneurysm but merely demonstrates a shift and stretching of vessels due to a space-occupying lesion, which may appear avascular. It has been suggested that most of these cases are due to hæmorrhage from a minute angioma which destroys itself in the process and is thus not demonstrable in routine pathological examination, but occasionally at operation a saccular aneurysm or its remains, may be exposed in the depths of the cavity from which the hæmatoma has been removed.

In most instances when the hæmatoma has been localized it can be drained by a simple burr-hole and needling, but if the contents are too solid to come easily through a cannula it may be necessary to turn down a bone flap, uncap the lesion and clear out the clot.

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THE SPINE AND SPINAL CORD

Injuries to the Spine and Cord

Fractures and fracture dislocations are nearly all produced by excessive flexion and so by indirect violence. The lower cervical and thoraco-lumbar regions of the spinal column are most often affected, the former by such accidents as falls on the flexed head, e.g. diving into shallow water or the collapse of the scrum on the football field; the latter by falls of heavy material, e.g. mine roofing or cases out of a cargo sling on to the shoulders of a miner or stevedore when bending forwards at his work.

Crush fractures of the vertebral bodies may produce few symptoms and little or no disability and are sometimes seen in miners and others when X-rayed for some other lesions, and who were quite unaware that at sometime in their past history they had fractured their spine (Rhys, 1934).

When it is remembered that the mechanism of production of fracture is flexion of the spinal column, the handling of an injured person who is suspected of having a fractured spine must be such that further flexion of the spine is avoided, lest an undamaged cord be injured or an incomplete lesion of it be converted into a complete one.

Injury to the cord is evident by either paraplegia or tetraplegia and if after 48 hours there is no sign of recovery (i.e. a period in which spinal shock may be regarded as having passed off) it is likely that irreparable damage has occurred and the case is that of complete or almost complete section at the level of the dislocation.

Hæmorrhage

Bleeding may be extramedullary or intramedullary and may complicate the clinical picture by producing a cord lesion at a higher level than the site of the dislocation. An

intramedullary hæmorrhage tends to spread longitudinally in the substance of the cord and so may reach a considerably higher level than the site of contusion.

The Level of the Lesion

Because of the difference in length of the spinal cord in relation to that of the bony column the segments in the cord are higher than the vertebræ to which they are related. A good working rule is to allow a discrepancy of one in the upper cervical, two in the lower cervical, three in the upper thoracic and four in the lower thoracic regions. Thus the tenth thoracic segment, the spinal nerve of which reaches the umbilicus, lies in relation to the sixth thoracic vertebral body.

The Treatment of Injuries

If the cord is not damaged the treatment usually falls to the orthopædic surgeon and consists of immobilization usually by the application of a plaster jacket. Early exercises to maintain the tone of the erector spinæ and abdominal muscles are encouraged while the patient is in the plaster cast.

For cervical lesions extension may be required by means of Crutchfield's tongs or some similar appliance.

If the cord is damaged, other considerations arise, the chief of which is whether laminectomy and exploration are likely to be helpful. Laminectomy has nothing to offer the patient with a complete lesion of the cord but may be indicated where there is only a partial injury. Any decision regarding operation must depend upon a conception of what surgical intervention may be expected to accomplish. Pressure on the cord whether as a result of fracture dislocation by indirect violence or as a result of direct violence, e.g. a blow or a missile, may be sustained. Unless it is obvious that the cord has been completely divided, laminectomy is indicated in such cases and also in the case of compound injuries, with the purpose of converting these into clean, closed fractures and at the same time freeing the cord from compression should such be present.

In other cases where spinal shock has passed off, i.e. after 48 hours, and there is evidence of some preservation of conduction, laminectomy is indicated if, (*a*) gross bony deformity is present or a missile is demonstrated radiologically in close proximity to the cord, (*b*) if there is severe root pain or, (*c*) if later an arrest takes place in what has been hitherto a progressive recovery. If doubt exists as to whether there is cord compression, one should investigate the subarachnoid space by lumbar puncture to ascertain whether or not there is any degree of spinal block present.

Spinal Block

The state of the subarachnoid space in relation to the flow of cerebrospinal fluid through it is ascertained by performing lumbar puncture with the patient lying in the lateral position. If there is any obstruction in the space the Queckenstedt phenomenon (rise and fall of pressure in the lumbar pond on compression and release of compression of the jugular veins at the root of the neck) is absent or modified. As the degree of spinal block increases, stagnation of fluid in the lumbar pond occurs with the consequence that the protein content of the fluid increases without alteration in its cellular content. Myelography not only localizes the block and its precise site (e.g. whether intradural or not), but also frequently indicates its nature.

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Injuries to the Spine and Cord

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TUMOURS OF THE SPINE

Extra-dural. These may occur in the bone or in the epidural space and may be primary or secondary. Secondary tumours are usually metastases from carcinoma of the breast,



× 100

FIG. 35. Microscopical appearance of metastatic spinal tumour from thyroid carcinoma

(From The Annals of the Royal College of Surgeons England)

thyroid, lung, prostate, testis, or adrenal and are clinically characterized by a rather sudden onset of paraplegia, rapidly becoming flaccid in its type, which occurs in a patient who is usually middle-aged and harbours a primary neoplasm of one of these types. (We may cite cases of a secondary thyroid tumour (Fig. 35) in the laminæ in the mid-thoracic region of a man aged 44, of adeno-carcinoma of the breast in the upper thoracic region of a woman aged 42 whose left breast had been removed for carcinoma nearly 3 years before, metastasis in the lower thoracic region of an elderly man from a seminoma of the testis, in another aged 58 from the prostate and in still another aged 54 from the adrenal. Another patient, a man aged 79 had a metastasis from carcinoma arising in a prostatic remnant after enucleation of the prostate 28 years before.)

Primary tumours of the vertebræ, whether benign or malignant are not common. The commonest are plasmacytomas (Fig. 36), either solitary or appearing as part of diffuse myelomatosis. They occur in the epidural space and compress the theca and cord. (We may cite a case of paraplegia produced by what was apparently a solitary plasmacytoma

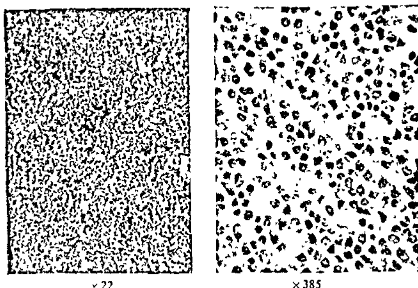


FIG 36 Typical microscopical appearance of plasmacytoma
(From The Annals of the Royal College of Surgeons, England)

in a man aged 47. He recovered from his paraplegia after laminectomy and removal of the neoplastic mass and lived some 5 years, but developed generalized myelomatosis from which he then succumbed.)

Neurinomas sometimes occur outside the dura and are then usually of more cellular type than those which are more commonly met with in connection with the intradural course of the nerve roots.

Lipomas may occur in the epidural space and compress the cord. Occasionally they have an intradural connection with corresponding intrathecal tumours. We have met one such case in a woman aged 34.

Tumours of the Membranes, Nerve Roots, and Cord

Tumours of the Cord. While metastases are not uncommon in the brain we have *not encountered one in the spinal cord In one instance an intrathecal metastatic deposit* lay outside the cord and was from a primary malignant adrenal tumour in a man aged 54.* True primary tumours of the cord itself are comparatively rare and are either gliomas such as ependymomas or hæmangioblastomas and rarely amenable to any form of treatment other than decompression and irradiation. Occasionally, however, the less malignant gliomas, e g. the ependymomas, may be dissected out of the substance of the cord with a good deal of benefit to the patient Elsberg's method of an incision longitudinally in the mid-line posteriorly made down to the tumour, then closure of the wound for some days and a second attack, is sometimes helpful, for at the second operation a certain amount of natural extrusion of the tumour may have occurred. This may facilitate complete removal of the tumour by opening up the way for its enucleation

* We have seen subarachnoid deposits of lymphadenoma and secondary squamous carcinoma from the cervix uteri.

from the substance of the cord. Very highly malignant ependymoblastomas and neuro-epitheliomas may occur in young subjects and are fortunately very rare.

Cysts in the Cord. Cysts in the cord are not uncommon, occurring chiefly in the cervico-thoracic region in syringomyelia but also in association with tumours such as ependymomas. Others are of obscure origin but patients with them have done well after the cysts have been drained into the subarachnoid space. If cysts in the cord are sufficiently large to obstruct the subarachnoid space, improvement in the patient's symptoms may be expected to follow decompression. One of our patients, a young woman aged 21,











FIG. 37. Meningiomas removed from women aged 55, 56, 75 and 78 respectively.

FIG. 38. Meningioma removed from a man aged 44.

(From The Annals of the Royal College of Surgeons, England)

has had extensive decompression for a diffuse cystic disease of the cord, the posterior arch of the atlas and all the spinal laminæ having been removed in 3 stages, as low as the fifth lumbar vertebra.

Membranes. Tumours of the membranes are either extra-dural or intra-dural, the commonest meningeal tumour being the meningioma (Figs. 37, 38) arising in the lepto-meninges and varying considerably in its degree of cellularity (Fig. 39). Some are more fibrous than others and many show the presence of psammoma bodies. Some contain bone (Fig. 40). They are among the commonest of spinal tumours and if detected early and removed before conduction in the cord or cauda equina is seriously impaired as a result of compression, the results of operative treatment are excellent (Fig. 41). They occur in both sexes but more commonly in middle-aged women than in men. Our youngest patient was a girl aged 16, our oldest a woman aged 78. An area of the related meninges, dura as well as arachnoid, should be removed along with these tumours so as to avoid recurrence from tumour cells which might otherwise remain in the field (Fig. 42).

Primary tumours of the vertebræ, whether benign or malignant are not common. The commonest are plasmacytomas (Fig. 36), either solitary or appearing as part of diffuse myelomatosis. They occur in the epidural space and compress the theca and cord. (We may cite a case of paraplegia produced by what was apparently a solitary plasmacytoma

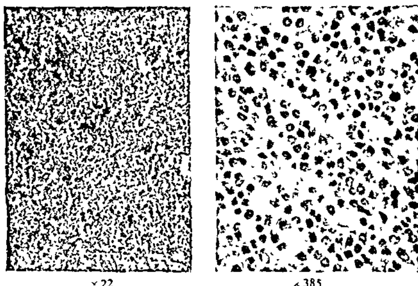


FIG 36 Typical microscopical appearance of plasmacytoma
(From The Annals of the Royal College of Surgeons, England)

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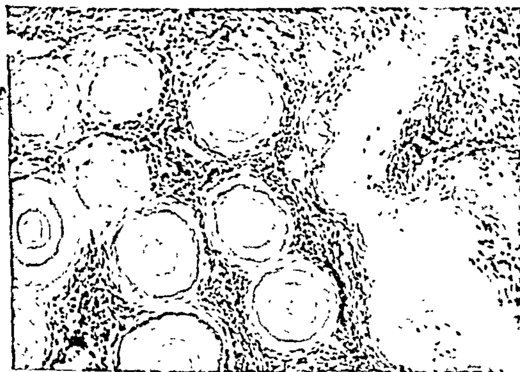
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* We have seen subarachnoid deposits of lymphadenoma and secondary squamous carcinoma from the cervix uteri.



(a)



×72 (b)

Fig. 40 Meningioma containing bone removed from a man aged 59.
(From The Annals of the Royal College of Surgeons, England)



187

FIG 39 Typical microscopical appearance of meningioma from a man aged 28 There are abundant psammoma bodies present

(From The Annals of the Royal College of Surgeons England)

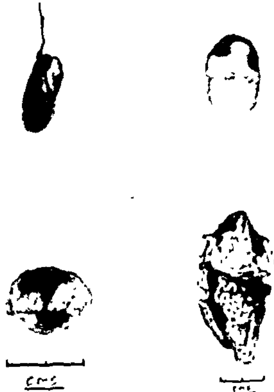


Fig. 43. Neurinomas removed from men aged 32, 33, 44 and 44 respectively.

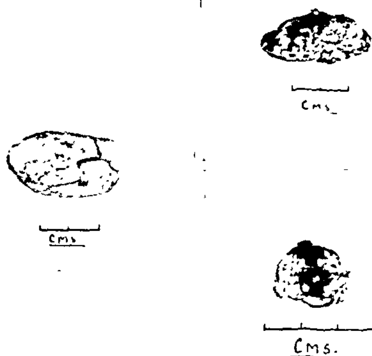


Fig. 44 Neurinomas removed from women aged 23, 50 and 62 respectively
(From The Annals of the Royal College of Surgeons, England)

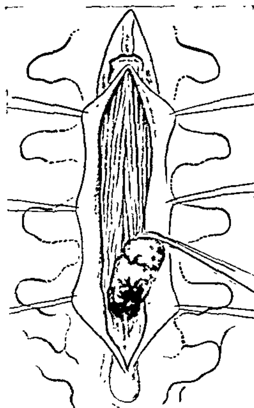


FIG 41 Removal of a cauda equina tumour.
(From "British Surgical Practice," Vol. 7, Butterworths)





FIG. 42. Meningioma with attached dura mater, removed from a woman aged 55.
(From The Annals of the Royal College of Surgeons, England)

straightened spine and narrowed intervertebral spaces sometimes seen in lesions of the intervertebral disks.

Myelography. The radio-opaque liquid may be introduced into the subarachnoid space either by cisternal or lumbar puncture. Myodil introduced into the lumbar pond and followed on the X-ray screen when the patient is tilted head or feet downwards, is the usual procedure at present. Where there is doubt as to whether or not there is a space-
[illegible], particularly helpful and may
[illegible]calize the lesion precisely and



Fig. 45. Dense (ivory) appearance of thoracic vertebra invaded by metastatic prostatic carcinoma in a man aged 58 with paraplegia from compression of the cord by the lesion.

(From The Annals of the Royal College of Surgeons, England)

perhaps also know its nature. Occasionally arachnoiditis has occurred after myelography and the investigation should therefore be undertaken only where the indications for it are definite.

Cervical Spondylosis and Intervertebral Disk Lesions

Many cases of persistent back-ache and sciatica are due to lesions of the intervertebral disks, particularly those separating the fourth and fifth lumbar vertebræ and the fifth lumbar vertebra and the sacrum. Operation in these cases is only indicated if conservative measures have been given fair trial without relieving the patient. Many patients recover with rest, either medical, i.e. in bed, or orthopædic, i.e. in a jacket; others are relieved by manipulation but this should only be undertaken if lumbar puncture has shown an absence of any degree of spinal block which if present is an indication for operation. When operation is undertaken a good result may be expected if the disk extrusion is

Epidermoid and dermoid tumours are rare and occur within the theca of young subjects, usually children.

Nerve Roots. The neurinoma arising from the nerve roots (Figs. 43, 44) is slightly more often encountered as an intrathecal tumour than is the meningioma and together these two tumours constitute the bulk of spinal tumours occurring within the dura. Like



FIG 45 Typical microscopical appearance of a spinal neurinoma This was a dumb-bell tumour from a woman aged 27

(From The Annals of the Royal College of Surgeons, England)

the meningioma, the neurinoma most often occurs in the thoracic region and is usually postero-lateral in its relationship to the cord. During its removal it is sometimes necessary to sacrifice the nerve root from which it arises and it should not be forgotten that an artery frequently accompanies the root. This vessel should be clipped or coagulated if it is necessary to divide the root with which it is associated.

Dumb-bell Tumours. These are usually neurinomas which, arising from the sheath of the issuing spinal nerve, have intraspinal and extraspinal parts connected by a narrow isthmus which lies in the intervertebral foramen through which it passes and which in the course of time it may enlarge considerably They are best dealt with by removing the spinal part of the tumour by laminectomy and at a later stage carrying out excision of the cervical, thoracic or abdominal part, according to the region of the spine in which the tumour occurs. The bulk of the intraforaminal part of the tumour is usually removed at the laminectomy and any remaining part of it when the extraspinal part is excised (Fig. 45). Although most commonly intrathecal these tumours are occasionally entirely extradural.

Radiology in Diagnosis

X-rays of the spine may very occasionally show the tumour as in one of our cases, a heavily calcified, intrathecal meningioma, more often changes in the bone are revealed, thinning or erosion of the pedicles, collapse or erosion of the vertebral bodies or heavy calcification of the body, as in metastatic carcinoma from the prostate in another of our cases (Fig. 46) X-rays also demonstrate the arthritic changes of spondylosis and the

more often it is only temporary but may last for from 6 months to 2 years. It may then need to be repeated or followed by operative treatment. This may consist of partial or complete division of the sensory root of the Gasserian ganglion or section of the trigeminal tract in the medulla. The operation most commonly performed is on the sensory root exposed by either an extradural or intradural route through the middle fossa by elevating the temporal lobe. If the extradural approach is used it is necessary to secure and divide the middle meningeal artery as it passes through the foramen spinosum. Corneal sensation may be preserved by dividing only the lateral two-thirds of the root. It is claimed that the root need not be divided but merely exposed and decompressed by widely opening Meckel's cave but we have no experience of this (Taarnhøj, 1952, 1954). The motor root which lies deep to the sensory and resembles a thin strand of white cotton, is preserved. The root may also be approached through the posterior fossa, as was advocated and practised by the late W. E. Dandy of Baltimore.

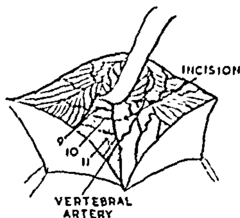


FIG. 47. Medullary tractotomy. (Sjorqvist's operation.)
(From the "British Medical Journal")

Medullary Tractotomy

The descending root of the trigeminus is divided by an incision in the brain stem, made between the origin of the vagal roots and the inferior olive. Sjorqvist (1938) (Fig. 47).

Neurotomy, Rhizotomy, Cordotomy, and Leucotomy for the Relief of Pain

Operative measures for the relief of intractable pain as in advanced malignant disease may be indicated when for any reason the continued administration of analgesic drugs is undesirable. These operative procedures comprise the severance of nerves or nerve roots or the pain pathways in the central nervous system, or their interruption or destruction by the injection of chemical substances such as procaine, alcohol, and phenol, electro-coagulation or suprasonic necrotization. The pain-conducting fibres in the spinal cord may be severed by a transverse section of the antero-lateral area of the cord in which they lie, or by a longitudinal section designed to divide them as they cross the midline as was suggested by the late Donald Armour at Queen Square. The operation usually performed is transverse section of the antero-lateral region of the cord after slightly rotating the cord by gently drawing on a slip of the dentate ligament (Figs. 48, 49). The knife is entered immediately anterior to the attachment of this ligament to the cord so as to make sure of not damaging the crossed pyramidal tract* (Kahn and Peet (1948); Kahn and Rand (1952)). Analgesia and thermoanæsthesia of the opposite side of the body are produced but the sense of light touch remains since fibres conveying it also travel in the posterior columns. The section must be made several segments above the level of the area in which the pain is felt because the upper limit of analgesia as

* The [illegible]
the rig[illegible]
on the [illegible] and obtained relief from them.

prominent and if after incising its coverings, a large mass of nuclear material extrudes and can be readily withdrawn from the disk space. There is no need to employ any form of spinal fusion or fixation subsequently.

Some relief from the effects of cervical spondylosis may be obtained from laminectomy and division of a series of dentate ligaments on each side of the cervical cord.

Where disk extrusions produce clinical pictures resembling tumours they are treated as such, i.e. by exploratory laminectomy.

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Tumours

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THE SURGICAL RELIEF OF PAIN

Pain is imponderable to the detached observer who can only approximately assess its severity by any objective manifestations in the subject of it. The same type of pain provoking stimulus must have a very different effect on a sensitive young woman from what it would on a hardened cattle-drover. In both it is but a symptom and its biological purpose for the most part protective. We must seek to allay its cause where this is possible. This may, however, be unknown or imperfectly understood, as in trigeminal neuralgia, or well known but no longer within the range of effective local treatment as in myocardial ischæmia or inoperable malignant disease. In such cases efforts must be wholly directed to relieving pain as a symptom. Surgical procedures should usually be considered only after lesser measures in the form of anodynes have failed or for any reason are no longer advisable. They comprise on the one hand the interruption of the pathways of painful impulses in their approach to the sensorium or within the brain itself. by such operations as neurotomy, rhizotomy, cordotomy, medullary or mesencephalic tractotomy or frontal lobotomy, or on the other, sympathectomy to increase the blood supply and thereby reduce or abolish the pain of ischæmia. Brief reference only, is here made to some of these procedures.

Trigeminal Neuralgia

True trigeminal neuralgia occurs characteristically in subjects over 50 years of age and affects the right side more than twice as commonly as the left It most frequently involves the mandibular and maxillary divisions of the nerve and is unassociated with sensory changes in the trigeminal field except perhaps in the direction of exaltation. The attacks are paroxysmal and the onset is frequently fired by a "trigger" such as the application of cold or the eating of something hard

Treatment

Alcohol injection of the Gasserian ganglion may give complete and lasting relief—

depressive and obsessional states which are chronic and progressive and for which all other measures of which we are cognisant have failed.

PAINFUL PHANTOM LIMB

After 4 years of age an amputation leaves a phantom or ghost of the limb which has been removed. In the course of time this gradually disappears but in some subjects the phantom persists and becomes painful, the lost part appearing to be distorted and sometimes magnified also, and subject to painful cramps. In attempting to relieve the sufferer of such a painful phantom various procedures have been carried out. Extirpation of parts of the parietal cortex has been undertaken but with disappointing results, but more recently the posterior columns in the cord conveying position sense have been divided with some degree of success (Pool, 1946; Browder and Gallagher, 1946, 1948).

Causalgia is a peculiarly unpleasant form of pain, varied by such conditions as temperature and the state of the weather, and affects the median or medial popliteal territory of some patients who have had amputations performed. These two nerves are peculiar in their blood supply and the vascular nature of the pain of causalgia appears to be well established. Repeated amputations which are often sought by the sufferers should be avoided. Sympathectomy relieves some, but occasionally spino-thalamic tractotomy (cordotomy) is necessary.

Prefrontal leucotomy does not abolish the perception of pain but may alleviate suffering by bringing about a lessened appreciation of pain as such, even if the operation is unilateral. The section need only be made in the medial ventral quadrant of the frontal lobe so as to divide the cortical connections of the thalamus while the neopallial lateral part of the lobe is preserved.

SURGERY FOR CONGENITAL AND DEGENERATIVE LESIONS OF THE NERVOUS SYSTEM AND FOR EPILEPSY

Surgical Treatment of such conditions as athetosis, hemiballismus (involuntary movements of an extremity) and Parkinsonism has been attempted, but the results have been disappointing. (*Vide* Surgery, Gynæcology and Obstetrics, 1955, 101, 488. An account of various procedures which have been performed in *the surgical treatment of epilepsy* will also be found in the same paper.) Numerous operations have been performed to try and arrest the fits but after they have been suppressed for a time they tend to recur whatever the procedure. If cortical scars are excised they tend to reform as do scars elsewhere.

Spina Bifida

This is a congenital defect in the bony coverings of the cord and its membranes and is often associated with protrusion of the contents of the spinal canal through the defect. If this is in the body of the vertebra the spina bifida is anterior, if in the neural arch, posterior. Posterior spina bifida is much the commoner and usually occurs in the lumbar region. The defect in the laminæ may be apparent only on X-ray examination or indicated only by a dimple, a pad of fat or a tuft of hair (spina bifida occulta). No treatment is called for unless symptoms arise due to traction on the cord by its fixation to the skin, when exploration is indicated. In gross forms there is a protrusion of spinal contents which may vary in degree and in extent. In myelocele the medullary groove is unclosed

demarcated immediately after operation may sink somewhat in the course of a few weeks. Sphincter disturbances follow the operation in some cases and though usually transient may persist, especially when the operation has been bilateral.

Psycho-surgery

The relationship between cerebral mechanisms and mind and personality is still unknown. In the American crow-bar case in which both frontal lobes were badly

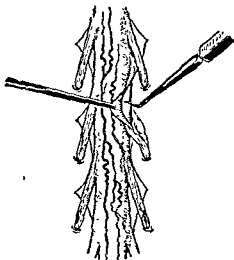


FIG 48 Spino-thalamic tractotomy (cordotomy) To expose the antero-lateral aspect of the cord it is slightly rotated by gentle traction on a slip of the dentate ligament.

(From the "British Medical Journal")

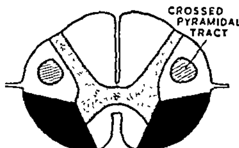


FIG. 49. Cross-section of spinal cord to show extent of area cut (black) in antero-lateral cordotomy.

(From the "British Medical Journal")

injured, a previously efficient foreman was said to have become weak, vacillating and profane and we have found that after removal of a cyst in the third ventricle a young boy who was violent and abusive and subject to rage attacks became docile and pleasant. Even extensive injury to the brain may have little or no detectable effect on personality, but changes of personality may be noticeable in subdural hæmatomas when there has been an accumulation of clot over a large part of one or both cerebral hemispheres. Frontal lobe lesions may produce euphoria comparable with the effects of alcohol, and some years ago Moniz injected alcohol into these lobes in mental patients, but subsequently abandoned this procedure for section of the lobes (lobotomy). The operation has been widely practised for various forms of mental disease and for intractable pain (*vide supra*) Originally the white matter was cut across in front of the anterior horn of the ventricle in each frontal lobe but various modifications of this simple section have since been practised. It does not abolish perception but alters its interpretation so that what was previously regarded as serious may now be taken lightly or even ignored. The section of the fronto-thalamic fibres however produces a personality change and the operation is being performed less often today than it was a few years ago. Its performance raises the [illegible]

are precise and the outlook for the patient otherwise bad. It may be of value in agitated,

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(rachischisis) in part or throughout its length. This condition is incompatible with any length of postnatal life and is not amenable to surgical treatment. The other gross varieties of spina bifida are meningoceles, meningo-myeloceles and syringomyeloceles.

Operation

This is contraindicated if there is pronounced neural deficiency or other associated and gross deformities. If there is no obvious neural defect operation should be undertaken to prevent leakage and meningitis and correct the deformity. Opinions differ as to the best time to operate but we favour either early intervention, as soon after birth as possible and find that these very young babies stand operation well, or else leaving them for 9 to 12 months. Not only is the early operation in the interests of the child but the mother is spared from seeing the unsightly deformity and carrying out daily dressings. When dissecting out the sac every effort must be made to preserve any nervous elements it may contain. The particular operation performed must be influenced by the type of spina bifida and the degree of its development but the essentials are the dissection of the sac to free it from its surroundings and after reducing its fluid content, its removal or plication so that it occupies the gap in the neural arches. Fascial flaps are then dissected up from the lumbar aponeurosis and used to cover in the spinal canal. No attempt is made to repair the bony defect. Further details of the surgical treatment of this and other spinal defects may be found in Grey Turner's *Modern Operative Surgery*, 12th Edition, 1956, pp. 486–492

Hydrocephalus

Infantile hydrocephalus is of two types designated obstructive and non-obstructive which may be differentiated by air studies and the tracing of phenolsulphonphthalein (phenol red) injected into the cerebral ventricles. Third ventriculostomy or the Torkildsen operation may be palliative in the obstructive variety. In non-obstructive hydrocephalus a number of operations have been performed such as ligation of both common carotid arteries, fulguration or excision of the choroid plexuses and drainage of cerebrospinal fluid into the ureter, the pleura, the retroperitoneal or intraperitoneal tissues. The mortality of these various procedures has been high and the results for the most part poor, but occasional successes are reported.

Infantile Hemiplegia and Extensive Unilateral Sturge-Kalisher-Weber's Disease (Associated Facial and Intracranial Hæmangiomata)

Improvement has been claimed to follow removal of the damaged hemisphere in young children the subject of birth injury of the brain or of this congenital vascular anomaly. It is claimed that convulsive attacks are abolished or lessened in frequency in about 80 per cent of cases and that there is an improvement in behaviour in many also. These advantages may outweigh the loss of visual field (a complete homonymous hemianopia) consequent on the hemispherectomy. After the operation speech is not usually affected irrespective of which is the dominant hemisphere (Carmichael, 1954).

References

Browder, E. J. and Gallagher, J. P. (1946) *N.Y. State J.M.* 46, 2403.
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since orbital injury is usually a part of a general facial injury, while injuries of the cranial cavity are mainly the responsibility of neurologists and neurosurgeons and, usually, only come under the care of the ophthalmologist for treatment of residual diplopia in that small percentage of cases in whom this complication does not settle spontaneously.

Injuries of the Eyeball

These injuries may be considered under the headings of: (1) Superficial injuries; (2) Perforating injuries; (3) Concussion injuries.

1. Superficial Injuries. Such injuries are the result of minor accidents causing small degrees of trauma. Corneal and subtarsal foreign bodies may cause shallow abrasions of the cornea and this type of lesion may also occur by scratches from a baby's finger or by branches of bushes in the garden or of trees in the countryside striking the eye unexpectedly. *Foreign bodies of the cornea* result from the entry into the eye of particles of dust or, in industry, from the passage into the eye of fragments of metal or other material from machines. It is the responsibility of the management of factories in which the risk of corneal foreign bodies occurs to provide protective goggles, but since there is no obligation upon the workers to wear them, the protection, frequently, remains unused. Corneal foreign bodies, inevitably, cause a small corneal abrasion which causes acute discomfort in the eye, and it is essential to avoid making this larger during the removal of the foreign body. Corneal foreign bodies may be of all sizes. The larger foreign bodies can be seen clearly, but the smaller ones can only be identified by the use of strong focal illumination and by the use of a magnifying lens (a loupe). A necessary preliminary to the *removal of a corneal foreign body* is the anæsthetisation of the cornea. This can be done by applying a drop of 4 per cent cocaine hydrochloride and repeating this two or three times at one minute intervals. Very superficial foreign bodies can be rubbed off the cornea with a small piece of cotton wool moistened in 4 per cent cocaine hydrochloride, but those which are situated more deeply in corneal tissue should be removed with a sharp pointed needle (Fig. 50). The spud is the traditional instrument for removing corneal foreign bodies, but it has a blunt end and causes increased abrasion of the corneal epithelium as compared with a needle, and should not therefore be employed. All corneal abrasions, whether the result of foreign bodies or of other trauma, heal rapidly if the eye is covered with a pad and a firm bandage. These abrasions may be associated with some intraocular inflammation called traumatic uveitis (page 101). Treatment involves the use of a cycloplegic and mydriatic drug which paralyses the ciliary muscles, thereby putting the eyeball at rest, and which dilates the pupil and prevents adhesions between the iris and the anterior capsule of the lens. All corneal abrasions, unless very small, should be treated with 1 per cent atropine sulphate drops or with 1 per cent hyoscine hydrobromide drops or with another similar drug, before the pad and bandage is applied. The main *complications* of a corneal abrasion are infection, which may lead to corneal ulceration and even to panophthalmitis (page 105), and recurrent abrasion, in which the symptoms of the abrasion recur days or weeks after it has apparently healed. The likelihood of infection is lessened if a drop of one of the antibiotic drugs such as penicillin 2,500 units per ml. or chloramphenicol ½ per cent is placed in the eye at the same time as the drop of mydriatic and before the pad and bandage is applied. Corneal abrasions must be kept under strict observation until healing is complete in order that infection may receive immediate treatment. Recurrent abrasion is caused, it is believed,

CHAPTER II

THE EYE

A. G. CROSS

SURGICAL procedures have, in earlier days of medical history, been performed on the eye by those surgeons who were responsible for all surgical procedures on the human body. The development of medical knowledge has led to specialization in the various branches of surgery, and ophthalmic surgery was one of the earliest divisions which was separated from the main subject and has come to be practised by surgeons who confine their activities to the study of the eye and of its diseases. Ophthalmology involves a knowledge of the pathological processes which affect the eyeball and the ocular adnexa, and of the manner in which general disease of the body may affect the eyes. It includes also an understanding of the normal and abnormal functions of those parts of the central nervous system which are responsible for the perception of vision, and for the movement of the eyes. An appreciation of the optical functions of the eyeball is necessary, and an understanding is required of the refractive faults which may result in defective vision or in eye-strain The methods of treatment of ocular disease, both medical and surgical, and of all optical abnormalities of the eyes must be understood by the ophthalmic specialist, but this knowledge is not necessary for the general surgical specialist, who should, however, be able to diagnose diseases of the eye which may occur as emergencies and to perform those surgical procedures which may be urgently required to preserve vision. He should also have a knowledge of ocular complications of those general surgical conditions with which he is regularly in contact. The most important surgical emergencies in ophthalmology are the treatment of injuries, the treatment of inflammatory conditions of the eye and ocular adnexa, and the relief of acute congestive glaucoma. It is true that the latter condition can often be relieved by conservative means, but sometimes surgical relief of the raised pressure is the only treatment which will save sight. The more important ocular complications of general surgical disease occur in association with tuberculosis and syphilis in their various manifestations, with arthritis and urethritis, with hypertension, with anæmia and leukæmia, with endocrine diseases causing exophthalmos, and with malignant tumours in many parts of the body, especially in the central nervous system.

OCULAR INJURIES

The eyes may be injured together with other parts of the body by explosions, by aircraft crashes and by traffic accidents, or the ocular injury may be the only damage. The eyes are a vulnerable part of the body and injuries to them may be more serious than would a similar injury on another part of the body A small abrasion of the skin may heal without obvious scar, while an abrasion of the cornea may become infected and cause the loss of the eye. Ocular injuries comprise injuries of the eyeball, and of the ocular adnexa, but the results of damage to ocular connections by injuries of the cranial cavity must also be considered. Injuries of the eyeball are usually the responsibility of the ophthalmologist, but injuries of the ocular adnexa are treated, in part, by the facio-maxillary surgeons

conjunctivæ, and cortisone drops which prevent that submucous inflammation of the conjunctiva which may lead to fibrosis and shrinking of the conjunctival fornices. Cortisone drops also minimize corneal vascularization during the process of repair so that, if subsequent keratoplasty (page 88) is indicated to improve visual acuity, this may be carried out with maximum chances of success. The injured eye must be kept covered with a pad and bandage and the use of all these drops continued until the corneal and conjunctival epithelium has healed.

2. Perforating Injuries. Perforating injuries of the eyeball may affect any part of the cornea or of the sclera, but, since the cornea is the anterior and unprotected portion, it or the adjacent sclera is most usually involved. The perforating injury of the outer coat of the eyeball is not important in itself. The serious aspect is the damage which may result within the eye, and especially to the crystalline lens and to the retina. Perforating injuries may be due to a large variety of causes: explosions both in war and peace, accidental stab wounds with pocket knives, scissors or screw drivers, firework injuries, air gun injuries, or the striking of the eye by small fragments of steel travelling at high velocity following the striking together of two larger pieces of steel. The knocking of a hammer against a chisel is, in civil practice, the commonest example of this latter type of injury. An injuring instrument passing through the cornea enters the anterior chamber of the eye which contains the aqueous humour. It may pass further to enter the lens and the vitreous body. Removal of the instrument from the anterior chamber results in a flow of aqueous out of the perforation, and this draws the flimsy iris into the opening, which it plugs so that no more aqueous can escape. Some small part of the iris probably lies outside the anterior chamber, and this condition is known as a prolapse of the iris. Perforating wounds which are caused by the entrance of small foreign bodies into the eye are not usually associated with any prolapse of the iris since it is the withdrawal of the perforating instrument which allows the outflow of aqueous humour and the sucking of the iris into the wound. Prolapse of the iris causes distortion of the pupil which is usually drawn towards the site of perforation. The iris may be injured by the perforating object, and the small hole which is made in the iris by a retained intraocular foreign body is often helpful in arriving at the diagnosis. Damage to the lens capsule results in some degree of lens opacity, and this is called traumatic cataract. Hæmorrhage from a torn iris may cause bleeding into the anterior chamber of the eye, the collection of blood in the anterior chamber being called a hyphæma. Perforations at the limbus, the zone where the cornea meets the sclera, may result in damage to the ciliary body, and sometimes the jelly-like vitreous body may appear in the wound. Larger wounds of the limbal region may be associated with complete extrusion of the lens.

Treatment of Perforating Injuries of the Eyeball

Small perforating injuries unassociated with a prolapse of the iris or with a retained intraocular foreign body may be treated by instillation of penicillin drops and 1 *per cent* atropine sulphate drops and by the application of a pad and bandage. Such eyes may have, or may subsequently develop, traumatic cataract, but the treatment of this condition is not urgent. The important indications are to make the wound heal rapidly and to prevent intraocular inflammation. Such patients should, if possible, be treated in hospital, but if this is impossible and they have to be treated at home they *must remain in* bed, except for toilet purposes. Continuous observation should be made in case infection

by the new and somewhat friable corneal epithelium becoming adherent to the tarsal conjunctiva during sleep. The act of waking and of opening the eyelid causes the friable corneal epithelium to be torn and a new abrasion is formed. This complication, which seems more likely to occur after scratches from fingers or from shrubs than after corneal foreign bodies, may occur repeatedly at intervals of a few weeks. It may be prevented by



FIG 50 Removal of corneal foreign body.

the instillation of some oily drop such as paroleine into the eye every night before going to sleep. Carefully graded applications of local ultraviolet light or of X-rays to the cornea are sometimes useful in preventing recurrences.

Local injuries of the conjunctiva are not of serious significance. Subconjunctival hæmorrhages rapidly absorb, and small tears of the conjunctiva can be sutured. *Generalized burns of the conjunctiva and cornea* by caustic fluids whether acids or alkalis may cause serious damage to the eye and permanent injury to vision. The superficial epithelium is destroyed, the cornea becomes partially opaque and the conjunctiva becomes raw. The corneal opacity may be permanent and the bulbar and tarsal conjunctiva may become adherent to each other. This is the condition of symblepharon, which results in inability to close the eyelids over the cornea, and, as a result, in further damage to that structure. *The immediate treatment* in all injuries of the eye by caustic fluids is to place the injured eye under a copious stream of water, as from a tap, in order to wash out all the residual caustic fluid. It is often stated that caustic acids should be washed out with weak alkali and that alkalis should be washed out with weak acid, but this is not important. A thorough washing with a bland fluid is the necessary treatment. Subsequently, it is necessary to instil 1 per cent atropine sulphate drops to combat the traumatic uveitis (page 101), oily drops such as paroleine to prevent adhesion of the bulbar and palpebral

of the lens, through the suspensory ligament, and then through a dilated pupil into the anterior chamber. They can then be removed through a keratome section. The vitreous cavity foreign bodies can also be removed by the so-called posterior route. A small opening is made through the conjunctiva, sclera, and choroid, 10 mm. or more behind the limbus, usually near to the position of the foreign body in the vitreous cavity. The magnet is then placed in this opening and the foreign body is removed. An application of diathermy should be made to the sclera at the site of the opening before the section is made, since this seals the choroid and retina to the sclera, and prevents retinal separation. It is helpful also to place a suture in the sclera before the opening is made so that the opening can be closed quickly as soon as the foreign body has been removed and before the vitreous body prolapses. Non-magnetic foreign bodies present a difficult problem. They must be accurately localized and then removed by the use of forceps inserted through a suitably placed incision in the eyeball. Radio-opaque foreign bodies can be localized by radiographical means, but others can only be localized by ophthalmoscopic observation, and since, in the presence of traumatic cataract, no view can be obtained, removal of some non-magnetic intraocular foreign bodies may not be possible.

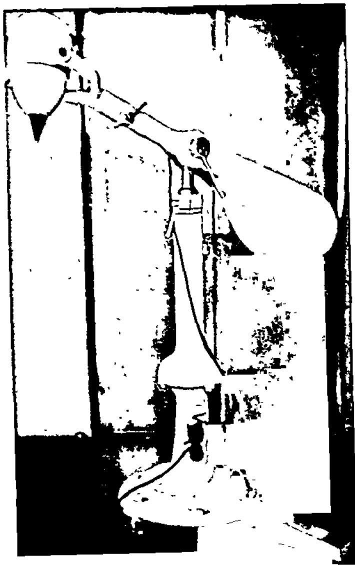


FIG 52. Philps' Giant Magnet.

Foreign bodies which penetrate the inside of the eyeball may carry infection, and the lens substance and vitreous form a suitable medium for the growth of bacteria. Generalized infection known as panophthalmitis may occur (page 105), usually due to one of the common pyogenic organisms, but occasionally due to Bacillus pyocyaneus or even to gas forming organisms like Clostridium welchii. Prevention of panophthalmitis can be accomplished most effectively by the systemic administration of antibiotics in full dosage. Penicillin is usually administered, and should be commenced as soon as possible after the entrance of the foreign body. The appearance of evidence of intraocular infection in the form of haziness of the aqueous, pus in the anterior chamber (hypopyon), pus at

should develop (page 105), as indicated by increasing conjunctival congestion, the presence of pus in the anterior chamber, the appearance of a yellow reflex in the pupil, and the loss of projection of light. Cases which have a large perforating wound, or which have prolapse of the iris, or which have a retained intraocular foreign body should be admitted to hospital in order that they may undergo surgical repair at the soonest possible moment. This procedure should, except under special circumstances, be carried out under a general anæsthetic. It can in an emergency be carried out using a local anæsthetic, the most satisfactory method being the injection of 1 ml. of 2 per cent procaine into Tenon's

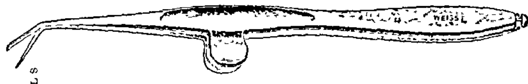


FIG. 51 de Wecker's iris scissors.

capsule, and the injection of 4 ml. of 2 per cent procaine around the facial nerve where it crosses the neck of the condyle of the mandible. Prolapsed iris should be grasped with iris forceps, withdrawn through the perforation until it is fairly taut, and cut close to the cornea with de Wecker's iris scissors (Fig. 51). The portion left will withdraw into the anterior chamber by its own elasticity, so that it lies clear of the corneal wound. The corneal wound must be repaired either by suturing the margins together with fine silk or by separating the conjunctiva at the limbus above or below, undermining it and drawing it down or up as a flap to cover and to splint the corneal wound. Conjunctival flaps can also be drawn across from the sides but they tend to be under greater tension in these situations and they retract more rapidly. The flap is held in position by two sutures, placed one on each side, fixing it near the limbus in the position required. Atropine sulphate drops and penicillin drops are instilled and a pad and bandage applied. The fixation sutures of the flap usually cut out, but, if they do not, they can be removed in a week and the flap will retract to its proper position at the limbus, leaving the wound healed and the anterior chamber well formed.

Removal of Intraocular Foreign Body

An intraocular foreign body must be removed from the eyeball, unless it is composed of some material which will not cause any reaction in the eyeball if it remains. Steel must always be removed because if it is retained in the eye it causes that condition of brown staining of the lens capsule, uveal tract, and retina called siderosis. Copper often causes a violent reaction resembling panophthalmitis and called chalcosis or chalyrosis, and it requires removal. Sometimes the copper particles may be deposited under the anterior lens capsule forming the typical "sunflower" appearance. Vegetable matter, such as wood, and also glass should be removed. Steel foreign bodies are removed with a magnet. This may be of the giant type, such as the Philps magnet (Fig 52), or sometimes a smaller hand magnet will be adequate. Those foreign bodies which are in the vitreous chamber may be removed by one of two methods They can be pulled to the equator

Removal of an Injured Eye

An eye which has suffered very severe injury as may occur in the explosions of war, may require immediate removal. Conservative treatment should be undertaken, however, if there seems the slightest chance that some degree of vision can be preserved. Some eyes which appear to be damaged beyond all possibility of recovery, heal satisfactorily and retain some amount of vision which, although not of great use, is better than no vision at all. The absence of light perception in an eye 6 or 7 days after the injury is a bad prognostic sign, and such eyes should be removed at that time since they may excite sympathetic inflammation in the other eye.

Sympathetic Ophthalmitis

This condition, although an inflammatory process, is considered in association with perforating injuries of the eyeball, because it demands continual consideration during the course of the treatment of wounds of the eye and because it is of such serious prognosis. It is an inflammation of the uveal tract (page 104), which affects the uninjured eye (sympathizing eye) as well as the injured eye (exciting eye), after a perforating wound of the eyeball, whether accidental or surgical, and which may persist in spite of all treatment until both eyes are so severely damaged that blindness ensues. Ophthalmic practice suggests that removal of the injured eye before the tenth day after the injury prevents the onset of sympathetic ophthalmitis in the uninjured eye, and this is the reason that eyes which have no possibility of recovery of vision should be removed at the end of the first week.

Wounds of the eyeball in the region of the limbus have more tendency to induce sympathetic ophthalmitis than those situated in the middle of the cornea or in the sclera behind the limbal region, possibly because, at this part of the eye, there is more chance of adhesions between the uveal tract and the wound, which may cause continued irritation. The onset of sympathetic ophthalmitis may be delayed for weeks, months, and occasionally for years after the injury. It seems true, however, that an eye which settles completely after a wound is unlikely to induce a subsequent sympathetic inflammation. The cause of sympathetic ophthalmitis is uncertain. The two theories are: (1) Spread of an infective organism, possibly a virus, from the injured to the uninjured eye, possibly along the optic nerve; (2) Inducement of a state of hypersensitivity in the uveal tract of the uninjured eye which renders it sensitive to toxic products originating in the uveal tract of the injured eye which are harmless to the rest of the body.

Rupture of the Eyeball

This may occur as the result of a blow in the eye from some blunt object such as a fist or a club. The rupture usually occurs circumferentially at the weakest part of the sclera which is about 3 or 4 mm. behind the limbus. Prolapse of the ciliary body and of the vitreous body, and extrusion of the lens, may occur. This is a serious injury and diagnosis may be delayed because, due to extensive bruising, the eyelids may be difficult to separate so that inspection of the eyeball is a matter of great difficulty. Repair of the wound must be effected by cutting off the prolapsed uveal tissue and vitreous and placing the margins of the wound in apposition by means of fine silk sutures. The subsequent treatment is similar to that of all perforating wounds. Rupture of the sclera without damage to the conjunctiva may occur, and, in these cases, subconjunctival dislocation of the lens may be found.

the margins of the wound of entry, or pus in the vitreous, indicates that other antibiotics should also be administered. Gas gangrene may be diagnosed by the presence of bubbles of gas in the anterior chamber of the eye, and should also be treated by the systemic application of antibiotic preparations. Uncontrolled intraocular infection may necessitate removal of the eye (page 133).

Recovery from perforating wounds of the eye may be slow, and it is influenced by the degree of damage to the crystalline lens. A perforating wound of the cornea, without damage to the lens, and which is properly treated by abscission of the prolapsed iris, and by closure of the rent, may heal in a few days with recovery of vision approaching the pre-accident standard. Damage to the lens, however, causes a different prognosis. The lens matter is gradually absorbed and it causes the eye to be irritable, injected, and watery. This may go on for a period of many weeks. The eye may settle when the lens matter has absorbed, but the irritation may be continued as the result of the adherence of small strands of lens capsule to the healed corneal wound. The vision will not be satisfactory because, even if the cornea and vitreous are clear and the pupil is not blocked by scarred fragments of lens capsule, the optics of the eyeball will have been disarranged and a thick convex lens in the spectacles will be necessary to obtain satisfactory visual acuity. The image seen through this lens will be larger by about one-third than that seen through the uninjured eye, and so double vision results. The injured person, in consequence, usually elects to make use of his uninjured eye and to neglect that which has been injured, and he becomes in many respects a one-eyed man. Binocular vision may be obtained, in some of these cases, by the use of a contact lens, and in suitable patients by the insertion of an acrylic lens into the eyeball to replace the crystalline lens which has been lost. This latter procedure is associated with some risk of further damage to the eye. Cases in which the lens capsule is not so severely damaged may settle down with an eye which is blind due to traumatic cataract, and in which the lens substance will not absorb. They may require surgical treatment for removal of the cataract, though since similar difficulties of binocular vision will occur this is not always carried out.

Keratoplasty

Injury to the cornea whether from foreign bodies, from abrasions, from perforating injuries or from caustic fluids results in scarring of the cornea and in loss of its transparency. Vision may be improved in these cases by the operation of keratoplasty or grafting of corneal tissue from a suitable donor. The donor material may be an eye which has had to be removed and which has a normal cornea or it may be from a cadaver. The eye, in the latter case, must be removed within 10 hours of death and must be preserved until required in liquid paraffin at a temperature of 4°C. It is not possible, in most cases, to preserve the cornea in this way for longer than 7 days. A circular portion of cornea is removed from the recipient's eye with a trephine of suitable diameter, and replaced by a piece of cornea from the donor, which is of similar size The grafted cornea is fixed by suitable sutures. The cornea which has been grafted remains clear in the majority of cases. Some patients have the corneal opacity completely or mainly in the superficial layers of the cornea. It is possible in these cases to pare off the superficial layers of the cornea, and to replace them by a similar lamella pared off the donor eye. This operation of lamellar grafting gives less satisfactory visual results, but it is a safer surgical procedure.

the form of a rosette. This type of cataract may retrogress after the injury but this is not dependent upon treatment.

DISLOCATION OF THE LENS (FIG. 54)

This may be partial or complete. Partial dislocation may occur in any direction, up or down, left or right, and it is diagnosed by observing the margin of the lens in one meridian, often only when the pupil is partially dilated. Complete dislocation of the lens

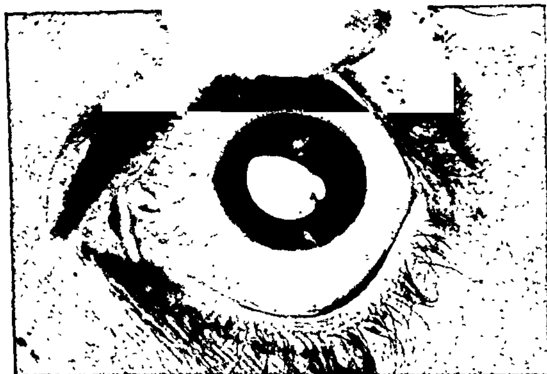


FIG. 54 Downward dislocation of the lens. (Stewart Duke-Elder.)

may be forward into the anterior chamber, where the lens can be seen with the naked eye, or back into the vitreous where it usually sinks to the bottom and can be seen with the ophthalmoscope. Dislocation of the lens causes visual disability, which may be overcome partially by suitable spectacles, though in complete posterior dislocation the eye is, for all practical purposes, aphakic and the difficulties of binocular vision already described will arise (page 88). The other complication of dislocation of the lens is secondary glaucoma (page 131), and this may necessitate removal of the lens, or some other surgical treatment. Removal of a dislocated lens can present many difficulties but it is not, usually, a matter of extreme urgency and full consideration can be given to the most suitable technique for the particular case before the operation is commenced.

COMMOTIO RETINÆ

This is an œdema of the retina which follows a blow on the front of the eyeball, and the damage takes place apparently by a contra-coup mechanism. The œdema affects particularly the layer of rods and cones, and when it has absorbed there may be some residual functional defect of that layer, associated with some proliferation of the pigment epithelium cells. This does not lead to serious visual disability unless the macular area is

3. Concussion Injuries of the Eyeball. These injuries are the result of blows on the eye with blunt objects, which are not of sufficient severity to cause rupture of the eyeball. They comprise hyphæma, iridodialysis and tears of the iris, concussion cataract, dislocation of the lens, commotio retinæ and rupture of the choroid.

Hyphæma

This is a collection of blood in the anterior chamber of the eye, which is often due to rupture of small iris vessels. The blood sinks to the bottom of the anterior chamber and

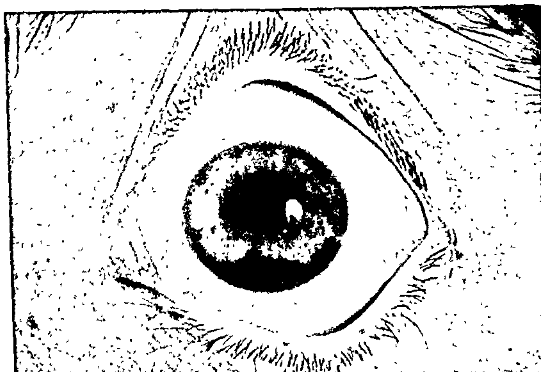


Fig 53 Traumatic iridodialysis (H Ridley)

gradually absorbs. Patients who present with this condition should be kept at rest in bed for 5–7 days because the bleeding may recur on the third or fourth day after the injury, and may be so profuse as to fill the anterior chamber, invade the vitreous and cause secondary glaucoma (page 131)

Iridodialysis (Fig. 53)

This is a tear of the iris at its attachment to the ciliary body, and it may be associated with hyphæma. Surgical repair of this lesion is possible, but since the condition does not cause any great visual disability it is only required for cosmetic reasons. Radial tears of the iris may also be caused by concussion injuries, and when incomplete they may affect only the fibres of the sphincter iridis muscle, causing a permanent partial dilatation of the pupil which is called traumatic mydriasis

Concussion Cataract

This is characterized by opacities in the posterior cortex of the lens which often have

Injuries of the Eyelids

These skin folds are very mobile and they move freely over the eyeballs in the process of blinking. They protect the surface of the cornea from small foreign bodies, and they spread over the cornea that tear fluid which is essential to its nutrition. Injuries of the eyelids may cause bruising, and they may involve damage to the surface or to the whole thickness of the structure.

Hæmatoma of the Eyelids

This is usually the result of a blow from some blunt object, and it is the traditional "black eye." The injury may have affected the eyelids or the bruising may be the result of the passage of blood from the scalp or forehead following an injury in that area. Swelling of the eyelids may be very great so that it is difficult to separate them to inspect the eyeball. Treatment requires the use of cold compresses to stop further bleeding. Absorption is usually rapid, and residual disability negligible.

Injuries of the Skin Surface of the Eyelids

These may take the form of small cuts or abrasions, or of burns. *Small cuts* may require suturing but they and abrasions usually heal rapidly in this area and without residual scarring. *Burns* may be the result of fire, caustics, or beta and gamma radiation and they may cause great disfigurement from scarring, while subsequent contracture of the eyelids may result in exposure of the cornea, and in serious corneal ulceration and opacity. The assessment of the prognosis in cases of burns of the eyelids necessitates an estimation of the depth of the burn. Superficial burns are characterized by a blister, and they usually heal without scarring. Deep burns cause total destruction of the epidermis, and show a white charred area of tissue which is slow to heal and which causes severe scarring and contracture. All burns cause general metabolic disturbance with considerable loss of plasma, but this is not likely to be a serious factor when small areas of facial skin are the only tissues involved. Patients with facial burns should be fully examined, and plasma transfusion should be given in all severe cases. Foreign bodies should be removed from the conjunctival sacs and oily drops such as paroleine should be instilled. Skin burns should be cleaned with 1 per cent cetrimide, and dusted with penicillin and sulphonamide powder. Crusts should be removed as they form and further powder applied. The hæmolytic streptococcus delays healing and causes destruction of skin grafts, but this organism is usually sensitive to penicillin. Staphylococcus is a common contaminant but, similarly, is sensitive to penicillin and does not usually retard healing. Wounds contaminated by strains of staphylococci which are insensitive to penicillin must be treated by other antibiotics. Superficial burns should heal in 14 days, but the deeper ones take longer. Sloughs separate and epithelium advances from the edges. The larger burns should be treated by skin grafts at the earliest opportunity because this hastens healing and lessens the tendency to contracture.

Scarring of the skin of the cheek and lower eyelid may lead to severe ectropion, and *grafting of skin* should be undertaken as soon as there is any tendency to this deformity. The principle of the operation is to incise along the length of the eyelid margin and to extend this incision beyond the inner and outer canthi. The incision is deepened and the skin undercut so as to give full mobility to the eyelids. A stent mould is made to fit the defect and this is covered with a sheet of Thiersch graft and fixed in position by sutures.

involved, in which case central vision may be distorted or defective. Physical rest is necessary after such an injury and this should persist until the absorption of the fluid is complete.

RUPTURE OF THE CHOROID (FIG. 55)

This may occur from the same type of injury as causes commotio retinæ. It usually occurs at the posterior pole of the eyeball, and often at the macula or circumferentially to

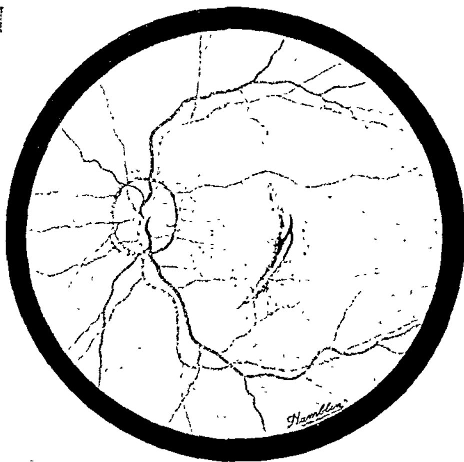


FIG 55. Ophthalmoscopic appearance of the fundus showing rupture of the choroid (Hamblin)

the disc. It appears as a whitish crescentic area with some pigmentary proliferation at its margins. The lesion may not be visible immediately after the injury since hæmorrhage may occur which spreads into the vitreous and prevents observation of the choroid and retina. The result of this lesion depends upon the part of the choroid affected. If the macula is injured, central vision is likely to be disturbed. Treatment consists of general rest and the use of 1 per cent atropine sulphate drops if signs of uveitis should develop. Concussion injuries of the eyeball may be associated with injuries of the eyelids, face, or head.

Injuries of the Ocular Adnexa

The eyelids and orbit afford protection to the eyeball, and they may be involved in injuries of the eyeball or they may be injured separately.

blunt objects. Fractures of the frontal or maxillary margins are not of serious importance, but a fracture of the malar bone on the lateral aspect of the orbit may cause damage to the orbital contents with associated double vision and it may require urgent treatment. This injury occurs from direct violence in motor cycle and cycle accidents, in football, in cricket, and in other activities. The bony fragment is displaced downwards and back and may penetrate the antrum which becomes filled with blood. The infraorbital nerve may be crushed causing anæsthesia of the upper lip, while displacement of the zygomatic arch may cause pressure on the coronoid process of the mandible and interference with movements of the jaw. Fractures of the malar bone should be elevated under general anæsthesia within 7 days of the accident, because the bone fragments become fixed and irreducible after that time. Fractures of the malar bone may be associated with fractures of other facial bones and these require appropriate treatment. Fracture of the maxilla may be associated with a fracture of the cribriform plate and with tearing of the dura matter. This is indicated by a leakage of cerebrospinal fluid from the nose and the loss of the sense of smell. Fractures of the ethmoid bone, which are the result of blows upon the face, may be symptomless until, following blowing of the nose, surgical emphysema of the orbit occurs causing protrusion of the eye. Chemotherapy should be given in these cases to prevent orbital cellulitis, and the patient should be warned not to blow his nose for 3–4 weeks.

Fracture of Face and Damage to Naso-lacrimal Duct

Fractures of the bones of the face may involve the naso-lacrimal duct and cause obstruction in this passage. Patients with these facial injuries may be acutely ill and frequently the obstructed duct is not diagnosed until, 3 or 4 weeks after the injury, there develops an attack of acute dacryocystitis.

Ocular Sequelæ of Head Injury

The eye is situated in close proximity to the brain and an injury of the cranium and of the cranial contents may affect the eye and the orbital structures directly, or, by damage to the nervous connections of the eyeball and its adnexa, it may cause indirect effects. The ocular sequelæ of head injuries may be classified as: (1) Signs which are important as a guide to general treatment; (2) Other immediate sequelæ; (3) Remote sequelæ.

1. Signs which are Important as a Guide to General Treatment

(a) The Pupils

Variation of the size and shape of the pupils of the eyes is common after head injuries. This may be the result of direct trauma to the eyeball, causing small ruptures of the pupillary margin of the iris, and the condition of traumatic mydriasis (page 90) or it may be the result of a lesion of the optic nerve, when the direct pupillary reaction to light is lost while the consensual remains, or of the oculomotor nerve. The pupils, in some patients, are observed to be normal immediately after the injury but subsequently one pupil dilates, and this may be followed by dilatation of a pupil of the opposite side. This sign is an indication of extradural or subdural hæmorrhage on the same side as the first dilated pupil.

(b) Deviation of the Head and Eyes

Deviation of the head and eyes towards the injured side is a useful localizing sign of a

Considerable overcorrection is necessary, because all grafts contract after they have been inserted during the process of healing.

Injuries Involving the Whole Thickness of the Eyelid

These may cause a considerable problem of plastic reconstruction. Simple cuts may be sutured, particular care being taken to obtain exact apposition at the eyelid margin and to cause some protrusion of tissue in this situation to allow for subsequent contraction. The conjunctival surface and the skin should be sutured separately and it is sometimes helpful to use a buried suture to hold together the middle layers of the eyelid. These wounds usually heal by first intention, but they may be infected from the conjunctival sac and breakdown occurs more commonly if the margins of the wound are forced together under tension. Loss of eyelid tissue may prevent the apposition of the edges of the wound and repair may involve grafting of mucous membrane to replace the conjunctiva and the use of a pedicle flap from the forehead to replace the skin. The eyelids are mobile structures, and after recovery from an extensive injury this mobility may be impaired, and result in some disfigurement. Injury of the inner end of the lower eyelid presents the added difficulty that the canaliculus is divided. It is important, when repair is being undertaken, that the two ends of the canaliculus are identified and that a style or a piece of silver wire is placed in them to hold them together during the process of repair. This routine allows the reformation of a patent canaliculus so that drainage of lacrimal secretion is not impaired.

Injuries of the Orbit

Injuries of the orbit may involve the soft tissues only or the bony walls. The soft tissues may be injured by explosions or by stab wounds from knives or scissors, or by falls on to pointed sticks which may sometimes traverse the optic foramen to damage the base of the brain. Foreign bodies, often of considerable size, may be retained in the orbit, without causing symptoms. A perforating injury of the soft tissues of the orbit may damage the *optic nerve* with resulting visual defect, or the oculomotor nerves and muscles may be traumatized causing defective mobility and double vision. Injury to the *ophthalmic artery* or to the *central artery of the retina* causes sudden blindness of that eye and a hæmatoma of the orbit may result. Leakage of cerebrospinal fluid indicates that the perforating wound has extended to the subarachnoid space. Introduction of infection may cause *orbital cellulitis* (page 107) and this may extend to cause *cavernous sinus thrombosis*. *Tetanus* of virulent form may follow perforating wounds of the orbit. *The treatment* of perforating injuries of the orbit requires full chemotherapy to prevent and overcome orbital cellulitis, the use of anti-tetanic serum, and the removal of retained foreign bodies Exploration for the removal of a retained foreign body may, in some cases, cause considerable damage to important structures and, if the foreign body is not causing reaction, it may be left undisturbed. Some foreign bodies can be removed from the front by an incision through the skin or conjunctiva, but others may require a lateral approach (Krönlein's method) or a superior approach (Naffziger's method).

Injuries of the bony walls of the orbit may result in communication between the orbit and the nasal sinuses or between the orbit and the anterior portion of the cranial cavity. Fractures of the bony margin of the orbit occur not uncommonly following blows from

palsy. The damage occurs at the time of the injury though diagnosis may be delayed. Paresis of one of the extraocular muscles results in compensatory changes in some of the other muscles. The majority of cases with damage to the oculomotor system recover spontaneously in the weeks or months which follow the accident, and only a small proportion require surgical treatment to overcome the residual double vision.

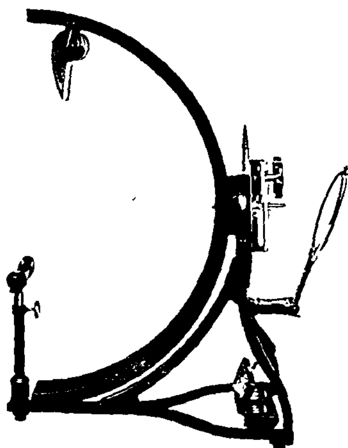


FIG 57. Lister perimeter. (Hamblin)

(c) DEFECTS OF OCULAR CONVERGENCE

This inability to turn both eyes inwards simultaneously is one of the most common ocular results of a head injury. It is due, in some cases, to oculomotor nerve defects, and in others it is associated with severe psychoneurotic manifestations. It appears to be, in the majority of patients, an indication of that general fatigueability which is a characteristic sequel of injury of the brain and it may cause some difficulty with reading and close work. It is easily overcome by regulated orthoptic training.

3. Remote Sequelæ

(a) ARTERIOVENOUS ANEURYSM

This vascular junction usually involves the internal carotid artery and the cavernous

destructive lesion of the base of the superior and middle frontal convolutions, provided the same convolutions are intact on the opposite side.

(*c*) PAPILLŒDEMA

This sign, though uncommon, does occur after head injury and indicates a rise of the intracranial pressure. It indicates that craniotomy may be required even although, at its first observation, the general condition of the patient is fairly good.



FIG. 56. Ophthalmoscopic appearance of optic disc with papillœdema. (Hansell.)

2. Other Immediate Sequelæ

(*a*) VISUAL TRACT

This may be damaged in any portion of its course, but the optic nerve and the optic radiations and visual cortex of the occipital lobe seem to be more vulnerable than the remainder of the visual nervous pathway. These lesions can be diagnosed by estimation of the visual fields on the perimeter (Fig. 57) and on the Bjerrum screen (Fig. 58). Partial recovery of the visual field loss may occur as the patient recovers from the effects of the injury, but some cases show a progressive deterioration which may seriously reduce the field of vision. Defects of the higher visual functions such as alexia, and visual object agnosia may also be found.

(*b*) OCULOMOTOR PALSIES

Lesions of the oculomotor muscles or of their nerve supplies are frequently found in association with head injury, the majority being an incomplete paresis and not a total

inflammatory reaction, in the absence of bacteria in the eyeball, by toxic products produced elsewhere in the body and to which it develops a hypersensitivity. This process affects chiefly the uveal tract, which comprises the corneal endothelium, the iris, the ciliary body, and the choroid, but the same reaction may also involve the cornea, the sclera, the retina, and even the optic nerve. The identification of the toxic substance is often difficult, and this type of inflammation may continue for long periods and may cause serious damage to the eyeball.

Uveitis is the most important intraocular inflammation and requires detailed consideration. It will be necessary subsequently to discuss panophthalmitis, a purulent inflammation of the whole eyeball, and endophthalmitis which is a non-purulent inflammatory reaction involving all the tissues of the eye. Orbital cellulitis, which is a generalized inflammation of the orbital tissues, and dacryocystitis, which is an inflammation of the lacrimal sac and its surrounding structures, will also be considered.

Uveitis

Inflammation of the uveal tract is one of the most common forms of ocular disease. The inflammation may affect, predominantly, the iris, the ciliary body, and corneal endothelium, and this is known as anterior uveitis (syn.: iritis: iridocyclitis), or it may affect, predominantly, the choroid when the condition is called posterior uveitis (syn.: choroiditis). The whole uveal tract may be equally affected by the inflammatory reaction which is then called panuveitis. Patients with anterior uveitis may show a reaction in the choroid, while patients with posterior uveitis may show some inflammation of the iris and of the ciliary body. The site of most marked inflammation determines the nomenclature. Uveitis may also be classified according to the acuity of the inflammatory condition, but this is not satisfactory since all gradations of acuity can occur between the two extremes, and, in most cases, the acuity of the reaction has no relation to the ætiological factor.

Pathology. The blood vessels of the uveal tract become dilated and an exudate of plasma-like fluid occurs into the tissue spaces. The tissue is infiltrated with inflammatory cells which are mostly lymphocytes, plasma cells and large mononuclear cells. Polymorphonuclear leucocytes occur usually only in response to pyogenic bacteria introduced through the blood stream or exogenously by perforating trauma. The inflammatory reaction may be generalized throughout the uveal tissues or it may be localized in the form of nodules. The nodular form is characterized by the continual healing of some nodules and by the development of new nodules which keep the ocular inflammation in a state of activity. Inflammatory cells and albuminous exudate circulate in the aqueous humour and the cells are laid down in clumps upon the posterior corneal surface: these are known as keratic precipitates or "K.P.," and since they can be observed on clinical examination they form an important diagnostic sign of uveitis. Inflammatory exudate from the ciliary body and from the choroid enters the vitreous and causes it to become hazy. Inflammation of the choroid inevitably involves the retina. The two membranes become adherent at the site of inflammation and at the margins of these adhesions there is hypertrophy of the pigment epithelium. An inflammatory exudate upon the surface of the iris may cause parts of its pupillary margin to adhere to the anterior capsule of the lens forming posterior synechiæ. These adhesions, if they extend all round the pupillary region, cause the anterior chamber of the eye to be separated from the posterior chamber

sinus, and it occurs weeks or months after the injury, and causes proptosis with œdema of the eyelids and of the conjunctiva. Pulsation of the eyeball may be visible. The condition is painful and corneal ulceration, and even perforation of the eyeball, may occur.

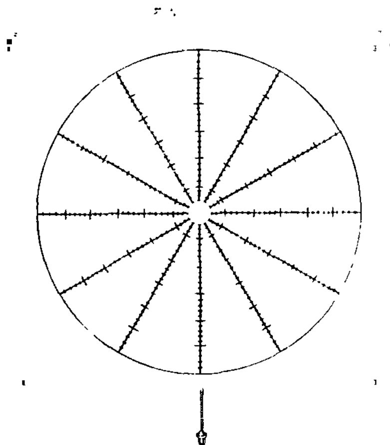


FIG. 58 Bjerrum screen. (Hamblin)

(*b*) ARACHNOIDITIS

A head injury is considered to be a predisposing cause in a proportion of persons who develop this condition of thickening of the arachnoid and pia mater. It leads to atrophy of nerve fibres and to visual defects.

INFLAMMATION OF THE EYEBALL AND OCULAR ADNEXA

Inflammations of the various parts of the eyeball and of the ocular adnexa are common conditions. The inflammation of that coat of the eyeball which is called the conjunctiva is one of the commonest of ocular diseases, and may be caused by many different bacterial organisms. It causes much morbidity in schools where it may occur in epidemic form, and among adults who may be prevented from attending their ordinary occupations. Localized inflammatory conditions occur on the eyelids as styes and as meibomian cysts and the orbit may become infected by pyogenic organisms to cause orbital cellulitis. These various forms of inflammation, caused by various bacteria, are similar in nature to the inflammation which occurs in all parts of the body, but the eyeball is a peculiar organ from the consideration of inflammatory processes. It may be affected by bacteria to cause local or general inflammation, but it can also, apparently, be stimulated to produce an

(4) TUBERCULOSIS. The uveitis may be due to the presence in the eye of Bacillus tuberculosis, or to the effect on the eye of tuberculous toxins produced elsewhere in the body.

(5) SARCOIDOSIS. This is a non-specific granulomatous inflammation which may involve any part of the body. The uveal tract is frequently affected.

(6) VENEREAL DISEASES. Gonorrhœa may be a cause of uveitis, usually as a sequel to a localized infection in the prostate gland. This type of uveitis which used to be common is less usually seen in these days, probably because the antibiotic drugs cure gonorrhœal infections very quickly. Generalized uveitis occurs in association with interstitial keratitis which is due to congenital syphilis, and uveitis may occur also in the secondary and tertiary stages of acquired syphilis.

(7) LEPROSY. Uveitis is a common finding in this disease during any part of its course.

(8) PRIMARY INFECTIONS OF ANIMALS WHICH ALSO AFFECT MAN. Brucellosis and toxoplasmosis.

(9) ARTHRITIS. Uveitis occurs frequently in association with arthritis. Still's disease of children is a rheumatoid type of arthritis frequently accompanied by uveitis. Spondylitis in adults is sometimes associated with very persistent uveitis.

(10) HERPES ZOSTER. This condition sometimes affects the ophthalmic division of the trigeminal nerve, and this may be accompanied by anterior uveitis.

(11) DIABETES MELLITUS.

(12) TRAUMA. Corneal abrasions may cause a uveal reaction and perforating wounds of the eyeball may lead to sympathetic ophthalmitis (page 104).

Symptoms and Signs. Pain is usually the first symptom of uveitis and it may be severe. It consists of a constant ache in the eye which may also affect the forehead and the upper jaw. Some patients, however, may suffer little or no pain. The eye may be congested and irritable, and watering may occur. Some visual defect is present as the result of the presence of inflammatory exudate in the aqueous humour and in the vitreous body. Floating specks may appear and the vision may become distorted. Defects in the field of vision may be noticed due to destruction of parts of the retina.

The most characteristic sign of active uveitis is the presence of a faint haze in the normally clear fluid aqueous of the anterior chamber. This is called the aqueous flare and it is due to the presence of inflammatory exudate in the aqueous humour. It may be observed by examination with focal illumination and the loupe in a dark room or by the corneal microscope. Œdema may be visible both in the epithelium and endothelium of the cornea. Striations may be found in the cornea due to folding of Descemet's membrane resulting from œdema, and keratic precipitates may be seen adherent to the posterior surface of the cornea. The iris loses its pattern and it becomes swollen and shows engorged vessels. The pupil is constricted until it is enlarged by the use of mydriatic drugs, and, if posterior synechiæ are present, it may be irregular in shape. The inflammatory exudate in the aqueous may be purulent and pus cells may sink to the bottom of the anterior chamber (hypopyon). Blood may be present (hyphæma), and in some patients a gelatinous film may collect in the anterior chamber. The eye is usually red and congested, especially in the circumciliary area, and the eyeball is tender on pressure. The intraocular pressure may be raised which is the condition of secondary glaucoma (page 131). The vitreous haze can be seen with the ophthalmoscope, and if this is not too dense

and is called seclusio pupillæ. The exudate may extend on to the anterior lens capsule in the pupillary region when, if it becomes organized, it causes the condition of occlusio pupilli. Continued secretion of aqueous humour when there is seclusio or occlusio pupilli leads to iris bombe, in which the iris is distended forward by the aqueous humour

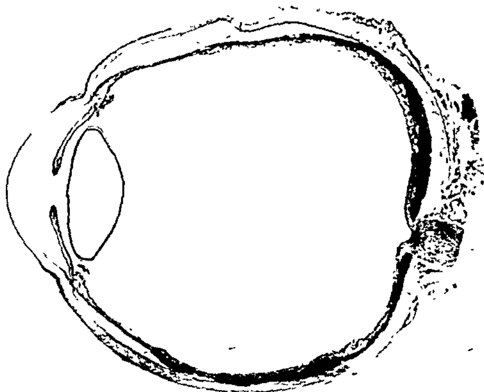


Fig 59 Sagittal section of eyeball with uveitis (N H. Ashton.)

which is unable to drain out of the eyeball. This causes obstruction of the angle of the anterior chamber and secondary glaucoma (page 131). Cessation of active inflammation of the uveal tract may be followed by absorption of all inflammatory products, or by organization. Some scarring remains and posterior synechiæ usually persist unless they have been broken down by treatment. Some cases of uveitis are characterized by the formation of nodules of inflammatory cells which are visible by clinical methods. This condition has been given the name of granulomatous uveitis, and it has been suggested that it is always due to the presence in the eye of some organism such as Bacillus tuberculosis, Spirochæta pallida, or Bacillus tularense. Clinical experience would not seem to confirm this opinion.

Ætiology. The possible causes of uveitis are many, but the most important can conveniently be classified as follows:

(1) A Focus of Infection in the Body. Infection of the teeth, nasal sinuses, tonsils, lungs, gall bladder, appendix, kidney, prostate gland, or other organs.

(2) Acute Specific Fevers. Measles, rubella, mumps, varicella, and variola may be characterized by uveitis in the early stage of generalized infection. Meningococcal meningitis may also show uveitis shortly after the onset of the illness.

(3) Specific Diseases of the Intestine. Typhoid, paratyphoid, and the dysenteries.

the regime adopted in a sanatorium for the treatment of pulmonary tuberculosis. Periods of physical rest with regulated exercise, accompanied by a nutritious, balanced diet, can cause considerable benefit. The good effects do not usually occur unless the patient can achieve absence of mental strain and of emotional trauma, and, since many patients are very unhappy when away from home for long periods, this treatment is not universally applicable.

(*a*) ADRENOCORTICAL PREPARATIONS. These endocrine preparations are of the greatest value in the treatment of uveitis. Cortisone, hydrocortisone, or other similar preparations, may be given systemically and cortisone and hydrocortisone can also be used locally to the eye (page 103). Systemic administration is indicated in the more severe cases of anterior uveitis, and in cases of posterior uveitis. This treatment is not, unfortunately, suitable for all patients. Hypertension and active pulmonary tuberculosis are absolute contraindications. The patient should take a salt-free diet while the preparation is being taken and the weight and blood pressure should be measured every 2 days. Any increase of weight or rise of blood pressure necessitates a reduction or a cessation of this form of therapy, though when possible this should be graduated and not sudden. Patients may be started on 100 mg. of cortisone or hydrocortisone a day and they may continue to take this for a week, and then reduce to 50 mg. a day for 2 weeks, and then to 25 mg. a day for a further 2 weeks. Marked increase of weight or a substantial rise of blood pressure is an indication that the dose should be halved, and further halved 3 days later. The therapy can then be stopped completely in a further 3 days. This treatment should, when possible, be given in hospital where all precautions can be taken to see that no complications occur. It is essentially suppressive rather than curative, but it appears that, if the condition can be suppressed, it is less likely to recur in such active form. Uveitis is, ultimately, self limiting, and if the eye can be maintained undamaged through the course of the inflammation, sight will be preserved. The adreno-cortico-tropic hormone of the pituitary gland (A.C.T.H.) may also be used in the treatment of uveitis.

(*b*) NON-SPECIFIC PROTEIN SHOCK THERAPY. This therapy may sometimes be beneficial in the treatment of uveitis. It seems likely that its action may be to increase the production of cortisone by the patient's adrenal cortex. Intravenous injections of T.A.B. vaccine may be used commencing with 25 million of the typhoid organisms, and increasing to 50 million, and 100 million as may be required on each third day. Alternatively, 5 ml. of milk may be injected intramuscularly and increased by 2 ml. at each dose on each third day, for 5 or 6 doses.

(*c*) TUBERCULIN. It has been considered that many of these cases are due to tuberculosis, and that they should be desensitized to tuberculin by repeated small doses of some preparation of tuberculin. Old tuberculin is often used commencing with 0·1 ml. of 1/100,000. This is increased by 0·1 ml. at weekly intervals up to 1·0 ml. One month's rest is followed by a course of 1/50,000 and the dosage gradually increased. One or two years' treatment may be required.

2. LOCAL

(*a*) ADRENOCORTICAL PREPARATIONS. These preparations may be used locally to the eye. Cortisone (2·5 mg. per ml.) and hydrocortisone (2·5 mg. per ml.) may be administered in the form of drops, and cortisone suspension (0·5 ml.) by subconjunctival injection. These applications give satisfactory effects in many cases of anterior uveitis, but do

white fluffy patches of inflammation can be seen contrasting with the normal red colour of the fundus. Settling of the inflammation is followed by absorption of the vitreous haze and by flattening of the patches of choroidal reaction. These areas are white as a result of the scleral tissue behind them and they have sharply circumscribed edges which may show some pigment proliferation.

Complications. The most frequent complication is *secondary glaucoma* (page 131). This occurs during the acute stage as a result of obstruction of the angle of the anterior chamber by fibrinous exudate, but it also occurs later in the disease as a result of seclusio and occlusio pupilli which, similarly, prevent adequate drainage of the aqueous from the anterior chamber of the eyeball. *Permanent corneal opacities* may occur as a result of organization of epithelial œdema, and *complicated cataract* occurs in some long standing cases of uveitis as a result of defective nourishment of the lens. Patients with posterior uveitis sometimes develop *optic neuritis* due to the spread of the inflammatory process from the choroid into the optic nerve, and in these cases the optic disc becomes blurred, the visual acuity becomes increasingly defective, and gaps may appear in the visual field.

Differential Diagnosis. Acute anterior uveitis may be mistaken for acute conjunctivitis or for acute congestive glaucoma (page 126). There should be no difficulty in making the differentiation if the facts in the following table are considered.

	Acute conjunctivitis	*Acute anterior uveitis*	*Acute congestive glaucoma*
History of Symptoms	Discomfort (not pain).	Pain, sometimes associated with looking at bright lights.	Acute pain, blurred vision, and presence of haloes around lights
	Conjunctival discharge with eyelids stuck on waking from sleep.	Blurred vision	
Signs	Congested eye	Congested eye.	Congested eye.
	Clear cornea	Deposits on posterior corneal surface.	Œdema of corneal epithelium
	Clear anterior chamber	Flare in anterior chamber	Shallow anterior chamber.
	Normal iris	Swollen, discoloured iris.	Congested, discoloured iris
	Normal pupil	Constricted pupil (unless mydriatic has been used).	Semidilated pupil.
	Normal ocular tension.	Normal ocular tension (unless secondary glaucoma has occurred).	Increased ocular tension

It must always be remembered, that some cases may present a dual diagnosis. Secondary glaucoma due to acute anterior uveitis may cause difficulties, but careful observation will always lead to an accurate diagnosis.

Treatment

1. General

The essential general treatment requires assessment of the cause of uveitis and treatment to eradicate this cause. Frequently, no cause is found. Many patients with persistent and prolonged uveitis are benefited by a course of treatment which is modelled on

but in doubtful cases removal is the safe procedure. Enucleation of the exciting eye has little effect upon the progress of the disease after sympathetic inflammation has developed in the sympathizing eye and treatment of such an established case is similar to that of all cases of uveitis. Many of these cases have an unfavourable prognosis and, as a result, many different forms of therapy have been used. The prescribing of sodium salicylate in large doses, salvarsan, and antihistamine drugs has been advocated, but it seems that they are unlikely to have any more beneficial effect than the routine methods already suggested.

Syndromes Associated with Uveitis

1. Uveo-parotid Syndrome (Heerfordt's Syndrome)

This condition consists of uveitis accompanied by bilateral swelling of the parotid glands, sometimes associated with facial palsy and occasionally accompanied by swelling of the lacrimal glands. It has a diverse ætiology and may be due to tuberculosis, sarcoidosis, and to other causes. Treatment of the ætiological factor and of the uveitis gives satisfactory results.

2. Behcet's Disease

Uveitis characterized by recurrent and rapidly absorbing hypopyon, together with ulceration of the mouth and of the genitalia. The cause of the condition is uncertain, but there is evidence that it may be a virus. The clinical course is steadily progressive and leads to blindness in all cases, usually as a result of damage to the optic nerve by recurrent optic neuritis. Damage to the ciliary body appears to be less than in some forms of uveitis and phthisis bulbi is not a usual sequel in these cases. Aphthous ulcers occur in all parts of the mouth, and superficial ulcers on the scrotum in the male and on the labia and clitoris in the female. Some patients may have erythema nodosum, arthritis and pyogenic lesions of the skin. Treatment of the eyes should be carried out according to routine for the uveitis, but it is rarely successful in preventing loss of sight.

3. Vogt-Koyanagi Syndrome

Uveitis associated with alopecia, vitiligo, poliosis, and dysacousis. Patients may be seen in whom the syndrome is not complete, and in whom one or more of the components are missing. It has been suggested that this condition is due to infection by a specific virus, but it is equally possible that it may have a multiple ætiology as has the uveoparotid syndrome. Treatment is according to the general principles already described.

4. Harada's Disease

Uveitis with retinal detachment and pleocytosis of the cerebrospinal fluid. The cause of this condition is unknown. It usually settles with reattachment of the retina and return of the vision to normal. The treatment of the uveitis is by the usual methods.

Panophthalmitis

An acute purulent inflammation of the whole eyeball, usually caused by pyogenic organisms carried to the eye in the bloodstream or introduced through a perforating

not have a marked effect upon inflammatory processes of the posterior segment of the eyeball.

(*b*) Mydriatic-cycloplegic Drugs. These drugs used as drops or ointment paralyse the sphincter pupillæ causing mydriasis and paralysis of the ciliary muscle. This results in palsy of accommodation and induces a state of rest for the eye. The dilatation of the pupil breaks down posterior synechiæ. These drugs may be instilled 3 times daily during acute phases of uveitis. One per cent atropine sulphate drops or 1 per cent atropine sulphate ointment are the mydriatics of choice, but patients who are sensitive to atropine may be treated by 1 per cent hyoscine hydrobromide drops, 1 per cent duboisine drops, 1 per cent eumydrin drops or 1 per cent lachesine drops. Subconjunctival injection of mydricaine, which is a mixture of atropine, cocaine, and adrenaline with boracic acid, gives a strong but transient mydriatic action, and is useful in breaking down posterior synechiæ in some cases. Fluid exudate into the iris tissue in cases of acute uveitis prevents full dilatation of the pupil during the active phase, and transient dilatation by mydricaine is always useful.

Treatment of Complications

1. Secondary Glaucoma (page 131)

2. Complicated Cataract

This may require extraction. The intracapsular method is most satisfactory and, if possible, should be delayed until all inflammation has subsided in the eye. Removal of cataract in an eye with subnormal tension is dangerous. Some degree of traumatic uveitis occurs in any intraocular operation, and, if the ciliary body has been seriously damaged by previous inflammation, the operation may cause failure of its function, further softening of the eye and phthisis bulbi.

3. Corneal Opacities

This may be treated by keratoplasty (page 88), provided the eyeball is otherwise healthy.

4. Optic Neuritis

This requires no special treatment; the essential therapy is similar to that for the uveitis.

5. Sympathetic Ophthalmitis (page 89)

Sympathetic inflammation usually begins insidiously and it is important to keep a close observation on the uninjured eye in all cases of ocular injury. Blurring of vision and some pain may be noticed by the patient, and an aqueous flare and keratic precipitates appear in the anterior chamber. Blurring of the optic disc may be an early sign since choroidal infiltration quickly extends backwards to the optic nerve head. Some cases may be characterized by a very marked amount of fibrinous exudate which causes much adhesion of the iris to the anterior lens capsule, and in these patients intraocular tension may rise for a time. In the terminal stages, however, the eye may become soft, and phthisis bulbi ensues. The most important treatment is prevention, which is carried out by removal of an injured and potentially exciting eye. A decision regarding enucleation is difficult if it is felt that there is a chance that vision may be regained in the damaged eye,

normal atmosphere. Mild degrees of retrolental fibroplasia may appear as a congenital retinal fold or as small patches of vitreous organization.

Treatment. These cases must be treated by the systemic administration of antibiotics, by rest, by atropine, and by heat. Enucleation is necessary in many cases.

Orbital Cellulitis

Infection of the contents of the orbit can occur from a variety of causes, and it leads to a generalized inflammatory reaction of the orbital tissues which may progress to suppuration. The infection may be introduced through the skin by a perforating injury or by surgical procedures on the orbit. It may also invade directly from the accessory sinuses of the nose through the thin bony wall which separates them from the orbit and particularly it may spread from the ethmoid group, or it may be carried to the orbit as a metastatic infection in the bloodstream.

Symptoms and Signs. The onset is associated with discomfort in the eye, and with aching pain in the orbit, and it usually progresses rapidly with increasing pain, swelling of the eyelid, protrusion of the eyeball, and limitation of movement. The fully developed case shows congestion and chemosis of the conjunctiva, marked proptosis, and almost complete absence of ocular movement. Pain may be very severe, especially in the cases which show suppurative changes and fever may be present. Vision is not, usually, seriously affected although in severe cases corneal ulceration, thrombotic changes in the ophthalmic artery or in the ophthalmic veins, or even panophthalmitis, may lead to loss of sight. Meningitis may be a complication. The infection can, also, extend back into the cavernous sinus to cause cavernous sinus thrombosis, and this is indicated by the onset of congestion and proptosis in the opposite eye.

Treatment. This necessitates the systemic use of one or more antibiotic drugs. Information regarding the susceptibility of the causal organisms is useful in deciding the preparation which should be employed. Infection of any of the nasal sinuses should receive surgical treatment, and, if severe pain suggests that suppuration is occurring, this may have to be drained by an incision through the upper or lower eyelid. Orbital cellulitis is a condition which, in the past, has sometimes been fatal, and has led to loss of vision in many patients. The danger to life was, partly, due to the complication of cavernous sinus thrombosis which was always fatal, but was sometimes the sequel to meningitis and to septicæmia. The introduction of antibiotic drugs has altered the prognosis, and this condition is not now considered a serious danger to life, although the vision of the eye may sometimes be destroyed in spite of all forms of treatment.

Dacryocystitis

Inflammation of the lacrimal sac is a common condition, and occurs in various forms: (1) Dacryocystitis of infants; (2) Chronic dacryocystitis; (3) Acute dacryocystitis.

1. The Dacryocystitis of Infants. This is due to the accumulation of cellular debris above an incompletely canalized naso-lacrimal duct. Discharge from the eye and regurgitation from the lacrimal sac is present from shortly after birth. Many of these cases settle spontaneously during the early months of life, and the regular emptying of the lacrimal sacs by pressure and the cleaning of the eyelids is the only treatment indicated at the onset. Persistence of the discharge is an indication for probing of the naso-lacrimal duct, and this treatment is usually completely and immediately successful.

wound, and which usually destroys it. The inflammatory cells which infiltrate all the ocular tissues, and especially the uveal tract, are predominantly polymorphonuclear leucocytes.

Symptoms and Signs. This condition usually follows a perforating wound of the eyeball, accidental or operative, but it may be associated with some general infection in the body, such as puerperal septicæmia. The onset is marked by severe pain in the eye, by complete loss of vision, and sometimes by such general symptoms as headache, vomiting, and rise of temperature. The eyeball is congested, the conjunctiva is œdematous, and the cornea may be hazy. The anterior chamber and the vitreous contain pus. The eyelids may be œdematous, and the eyeball protuberant, and perforation of the eyeball may occur in the later stages. Loss of ability to state the direction from which a light is shone into the eye is an indication that the retina has separated and that satisfactory vision will not be regained. The prognosis for recovery of vision in this condition is generally unsatisfactory, but it is agreed that purulent infection of an eyeball even after a perforating injury does not lead to sympathetic ophthalmitis. Panophthalmitis may be caused by an infection with Clostridium welchii, and gas bubbles may be found within the eyeball.

Treatment. The early treatment of all patients with perforating wounds by antibiotic drugs administered systemically is the most important factor in preventing panophthalmitis. Similar treatment after the condition has developed may be successful, though not invariably, and when the condition advances to the state when accurate projection of light is impossible, evisceration of the eyeball (page 135) may be required. Enucleation (page 133) is contraindicated because there is a danger that infection may spread along the subarachnoid space to cause meningitis. Evisceration or enucleation of the eyeball seems equally effective in cases of panophthalmitis which are caused by infection with Clostridium welchii. Panophthalmitis may sometimes follow infection of a corneal wound with B. pyocyaneus, and these cases do not respond to penicillin, but can be treated satisfactorily by streptomycin systemically, by subconjunctival polymyxin, and by polyfax ointment.

Endophthalmitis

A less acute generalized infection of the eyeball characterized by lymphocytic infiltration, but one which can be equally as destructive as panophthalmitis. The uveal tract is infiltrated with round cells, and the infection spreads to the vitreous. A zone of exudate may be deposited between the pars plana of the ciliary body and the posterior surface of the lens. This is invaded by fibroblasts to form a cyclitic membrane which may lead to retinal separation and to shrinking of the eyeball.

Symptoms and Signs. Irritation of the eye occurs following either a perforating injury or some general disease. Flare may be present in the aqueous and keratic precipitates are found. The vitreous becomes cloudy and the retina separates and travels forward to appear as a white mass behind the lens. The intraocular tension falls and, in due course, the eyeball shrinks. Infants sometimes show this condition soon after birth, and it is thought that it is the result of an intrauterine infection. It must be distinguished from retrolental fibroplasia, a condition which develops in premature babies during the first few weeks of life, and is thought to be the result of their being placed in a too high concentration of oxygen in the oxygen tent after birth and then being moved into the

intratracheal tube and to pack the pharynx. In either case the nasal cavity on the side of the operation should be packed with ribbon gauze which has been soaked in a mixture of 4 per cent cocaine hydrochloride and 1:1,000 adrenaline. The incision is about 1½ in. long and passes midway between the midline of the crest of the nose and the inner canthus of the eye, curving outwards below. The incision is made down to the periosteum and this is detached from the bone by means of a raspatory. The internal tarsal ligament is divided and all tissues, including the lacrimal sac, are retracted laterally to expose the lacrimal fossa. The bone at the bottom of the lacrimal fossa, which forms a portion of the lateral wall of the nose, is thin and is easily perforated with a Traquair's bone elevator. The opening is enlarged to a diameter of about ½ in. with punch forceps, care being taken not to damage the nasal mucosa. A longitudinal incision is made in the nasal mucous membrane and two flaps are formed by making two horizontal incisions. Corresponding flaps are made in the lacrimal sac and the anastomosis of the lacrimal sac is performed by suturing the two posterior flaps, and the two anterior flaps. No deep sutures are required before the skin is closed. A firm pad and bandage are applied for 24 hours. The skin sutures are removed on the fifth day, a lacrimal syringe is inserted, at this time, to the superior or inferior canaliculus and the new anastomosis is syringed through to demonstrate its patency.

2. Dacryocystectomy

This operation is usually carried out under local anæsthesia, 2 per cent procaine being injected around the lacrimal sac, but it can be performed under a general anæsthesia, when it is advisable to pass a tracheal tube and to pack the pharynx. An incision is made to the inner side of the eye starting above the internal palpebral ligament and passing down and out for ¾ in. The incision passes down to the crest of the lacrimal bone and a lacrimal sac retractor is inserted. The sac is detached from the bone medially, and then separated from the soft tissues above and laterally. It is severed below at the end of the naso-lacrimal duct. The duct is curetted as far as its entry to the nose, and the skin sutured. A pad, with pressure, is applied for 24 hours, and the sutures are removed on the fifth day.

OCULAR TUMOURS

The tumours which occur in the eye and in the ocular adnexa are, with some exceptions, similar to the tumours which occur in other parts of the body. There are a few which are peculiar to this region and there are some which behave differently when they occur in the eye, either as a result of anatomical or of other factors. Tumours which occur in the eye are often diagnosed early since either they are directly visible or they cause a visual defect which directs investigation to them. The prognosis, for this reason, may be better than when they occur in other organs of the body. Tumours of the eyeball can be removed by enucleation, and tumours of the eyelids can easily be excised, though the subsequent repair may present difficult problems for the plastic surgeon. Orbital tumours can often be removed satisfactorily, but those which involve the walls of the orbit cause difficulty and excision involves much mutilation. Treatment by radiotherapy is therefore of great importance in many of these patients.

2. Chronic Dacryocystitis. This usually occurs in adults, and it is most common in early middle age, and in women. The walls of the lacrimal sac become infiltrated with inflammatory cells and fibrosis follows. A stenosis occurs of the upper end of the naso-lacrimal duct and this causes a cessation of drainage of the tears from the eye, and an accumulation of pus and mucus in the lacrimal sac. The patient complains of watering of the eye, and pressure over the lacrimal sac causes a regurgitation of muco-pus into the conjunctival sac. This discharge from the lacrimal sac is infected, frequently with pneumococci, and if a small abrasion of the cornea occurs, this may also become infected causing the formation of a corneal ulcer which may progress to panophthalmitis. The presence of an obstruction in the naso-lacrimal duct may be confirmed by passing a lacrimal syringe along the canaliculus into the lacrimal sac and gently syringing. The passage of fluid into the nose and naso-pharynx is an indication of patency of the duct. Regurgitation of the fluid through the upper or lower canaliculi proves that the naso-lacrimal duct is obstructed. The important danger in cases of chronic dacryocystitis is that of corneal infection from the infected contents of the lacrimal sac. Many patients complain of serious discomfort from the continued watering and discharge.

Treatment. This may be of two types. Removal of the lacrimal sac (dacryocystectomy) removes both the focus of infection and the cavity where the muco-purulent discharge collects, but the patient is left, in most cases, with a perpetual watering eye. Dacryocystorhinostomy is a more satisfactory, though slightly more severe operation. An opening is made in the bony wall of the nose, the lacrimal sac is opened on its medial side, and the flaps of its walls are sutured to flaps of nasal mucous membrane. Drainage of tears takes place along the canaliculi through the lacrimal sac and into the nose. Probing of the naso-lacrimal duct in cases of chronic dacryocystitis is rarely effective. The mucous membrane tube is contained within a bony cylinder and it is impossible to dilate any stricture sufficiently to cause any permanent patency. Chronic dacryocystitis may sometimes be tuberculous in origin due to a spread of infection up the naso-lacrimal duct from lupus of the nose, and such cases must be treated by removal of the lacrimal sac.

3. Acute Dacryocystitis. This occurs as a result of obstruction of the naso-lacrimal duct frequently as a sequel to chronic dacryocystitis. The infection spreads to the tissues outside the walls of the lacrimal sac and a tense painful swelling occurs between the nose and the eye, below the internal palpebral ligament. Suppuration may occur and the pus points immediately below this ligament.

Treatment. This should be by antibiotics and by heat applications of all kinds. An abscess should be opened. It is almost unknown for orbital cellulitis or cavernous sinus thrombosis to occur as a complication of acute dacryocystitis. The condition tends to recur unless the obstruction of the naso-lacrimal duct is relieved by the operation of dacryocystorhinostomy, or the lacrimal sac is removed, and one of these procedures should be undertaken as soon as the acute inflammation has subsided.

Operations on the Lacrimal Sac

1. Dacryocystorhinostomy

This operation may be done through the nose (West's operation) or externally (Toti's operation). The external operation can be carried out either under a local anæsthetic or under a general anæsthetic, but in the latter case it is necessary to pass an

is common. General dissemination of the tumour may follow but sometimes it is long delayed.

Melanosis of the conjunctiva, an increasing subconjunctival pigmentation, may occur in fairly young persons. This condition is of serious prognosis if it is progressing. There is evidence that exenteration with removal of all the conjunctiva is the treatment of choice. The decision to undertake this procedure is difficult because the eyeball is often normal and has full vision, but it may be the correct treatment to save life.

(*d*) LYMPHOMA. Lymphatic deposits may occur in the subconjunctival region. Their exact nature may be difficult to define. Some are associated with lymphatic leukæmia, and some with lymphosarcoma, but others seem to be isolated deposits and all are treated satisfactorily by radiotherapy.

Uveal Tract

1. BENIGN TUMOURS

(*a*) ANGIOMA. This tumour is not seen frequently in the iris and ciliary body, but it is found in the choroid, when it may be associated with buphthalmos (infantile glaucoma). An angioma of the face, associated with an angioma of the choroid, makes up the Sturge-Weber syndrome.

(*b*) NEUROFIBROMA. This tumour may be found in the choroid, in association with neurofibromatosis of the eyelids, of the orbit or of the temporal fossa. Neurofibromatosis may be present in other parts of the body and cafe au lait spots or angiomata may be present.

(*c*) SIMPLE MELANOMA. This tumour is common in the iris and in the choroid. It has a similar structure to these tumours in the conjunctiva.

2. MALIGNANT TUMOURS

(*a*) MALIGNANT MELANOMA. The most common intraocular tumour is the malignant melanoma, and it occurs in the iris, in the ciliary body, and in the choroid.

Pathology. It is composed of polyhedral or of spindle-shaped cells, or of a mixture of the two, and it may be non-pigmented or very heavily pigmented, or there may be any intermediate gradation of pigmentation. Non-pigmented malignant melanomata are very unusual in the iris and ciliary body. The vascular supply of these tumours is very profuse, and the normal structure of blood vessels may be absent, only the endothelium being present to guide the blood among the malignant cells, and sometimes even this endothelial lining may be absent. Failure of the blood supply may give rise to zones of necrosis. These tumours contain varying amounts of reticulin which can be demonstrated by silver stains, and the amount of this may determine the dissemination of the growth, and be responsible, therefore, for the prognosis. Tumours with much reticulin seem to be less malignant than those with less reticulin. Choroidal malignant melanomata develop as flat growths, but they soon break through the membrane of Bruch at one small area, and the tumour seems to "pour" through this small area and to proliferate rapidly in the subretinal space, forming a mushroom structure.

Symptoms and Signs. Four stages of the development of intraocular tumours are described, and malignant melanomata develop in accordance with these stages: (1) *Stage of symptomless growth*: The tumour grows steadily but causes no symptoms. A tumour which can be seen will be observed to be increasing in size. (2) *Stage of raised intraocular*

Tumours of the Eyeball

Conjunctiva

1. Benign Tumours

(*a*) Papilloma. This may occur anywhere on the conjunctiva and it shows the characteristic structure of a vascular core covered by layers of stratified squamous epithelium. Treatment consists of removal of the tumour.

(*b*) Dermolipoma. This occurs on the outer side of the eyeball and may extend far back into the orbit between the extrinsic ocular muscles. It has the appearance of a fatty tumour and is closely adherent to the conjunctiva. It is composed of fat, and contains such skin structures associated with stratified squamous epithelium as hair follicles and sebaceous glands. These tumours are not malignant and their removal is not a matter of urgency. This is required sometimes for cosmetic reasons, and it is essential to avoid damage to the extrinsic ocular muscles.

(*c*) Hæmangioma and Lymphangioma. These occur in the subconjunctival tissue, but they show no particular characteristic in this situation.

(*d*) Simple Melanoma (Nævus). This may be pigmented or non-pigmented and it is usually situated near the limbus. It contains round cells with large nuclei (nævus cells), which, it seems, are derived from the epithelium. These tumours are usually situated near the limbus, but they do not usually increase in size, and so long as there is no evidence of progression it is not necessary to remove them. Increase in size is an indication for radical removal because it may be the result of the onset of malignant changes.

2. Malignant Tumours

(*a*) Rodent Ulcer. This may occur anywhere in the conjunctiva, but it usually appears near the caruncle. The structure is the same as when it occurs in the skin. There are columns of epithelial cells, the outer layer of which is columnar in type, and is called the palisade layer. Cystic spaces of varying size may be present. The tumour, in this situation, is a pink, ulcerated swelling which is locally malignant, but which does not disseminate. Treatment is either by excision or by radiotherapy.

(*b*) Epithelioma This tumour occurs, usually, at the limbus. Chronic epithelial hyperplasia of the conjunctiva, which is a precancerous condition, may precede the development of the malignant growth. An epithelioma has the same structure on the conjunctiva as elsewhere, but keratinization is less. The epithelium shows extensive proliferation, but cell nests may not be so frequent as elsewhere. The malignant cells invade the basement membrane, but they do not advance deeply into the fibrous tissue of the sclera and cornea. A limbal epithelioma appears as a pink ulcerated swelling, and the regional glands may be invaded. The treatment is removal of the eyeball.

(*c*) Malignant Melanoma. This may or may not commence in a simple melanoma, and it is composed of polyhedral or spindle cells, or a mixture of these cells, and all show many mitotic figures. The blood supply is often defective and may consist only of vascular channels lined with endothelium passing among the cells, and necrosis of clumps of cells may occur. Melanin may be present in these tumours which may grow rapidly and become pedunculated. Treatment may be removal of the eyeball with a large area of conjunctiva, but sometimes exenteration of the orbit may be necessary. Local removal may be justifiable in the case of very elderly patients, or with an only eye, but recurrence

detachment where the retina and retinal vessels show the folds of a simple separation. These tumours may, especially in the early stages, be very localized so that an area of normal fundus can be seen peripheral to them. Such an appearance never occurs in simple detachment of the retina, which always extends to the periphery of the fundus.



FIG. 61. Malignant melanoma of the iris.

Differential Diagnosis. The diagnosis of malignant melanomata of the iris does not cause serious difficulty, and the diagnosis of these tumours in the ciliary body can usually be made with certainty. The diagnosis of diktioma or epithelioma of the ciliary body must always be considered when non-pigmented tumours of the ciliary body are observed. The malignant melanoma of the choroid must be differentiated from a simple separation of the retina and from a simple melanoma. The simple separation of the retina does not, when viewed by the ophthalmoscope, have the solid appearance of a malignant melanoma, and when it is transilluminated by pressing a small light against the conjunctiva and sclera in the region of the separation, the glow in the pupil is clearly seen. A malignant melanoma, on the contrary, may show no glow when this is done. The presence of a retinal tear is evidence in favour of a simple separation. Simple melanomata are small, usually not more than three disc diameters in width, little raised, and unassociated with any simple separation.

Treatment. This should be by immediate enucleation of the eye. Careful examination, microscopic in addition to macroscopic, should be made to decide if extra ocular extension has occurred. Prophylactic irradiation may be given when this has occurred or in cases of doubt.

Prognosis. Death occurs by general dissemination in 30 per cent of cases of ocular malignant melanoma in 5 years, the organs particularly affected being the liver and the

pressure: This form of secondary glaucoma is common, *but the method by which the* intraocular pressure is raised is uncertain. (3) *Stage of extraocular extension*: The tumour invades the sclera and grows through it to proliferate outside the eyeball. (4) *Stage of general dissemination*: Tumour cells spread in the blood stream and form metastases

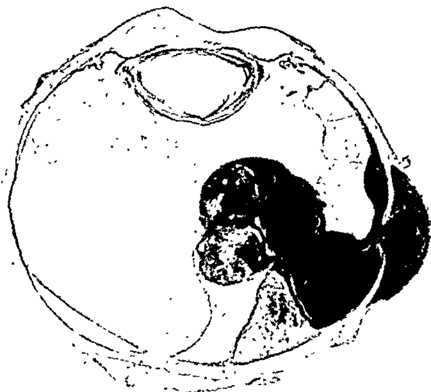


FIG 60 Sagittal section of the eyeball showing a malignant melanoma of the choroid, with extraocular extension (N H Ashton.)

elsewhere in the body. These stages are not necessarily consecutive, and they may happen simultaneously so that general dissemination may be occurring at the same time as symptomless growth.

Melanomata of the iris can be seen through the whole stage of their development. They may commence in simple melanomata and it may be difficult to decide when a malignant change has occurred. Progressive increase in size, and distortion of the pupil are significant signs in making a decision. Melanomata of the ciliary body may not be observed in their early stages, but as they grow they appear either in the angle of the anterior chamber, where they can be seen by the naked eye, or behind the lens where they can be observed with the ophthalmoscope. Malignant melanomata of the choroid may cause visual disturbances in the early stages, and they may, when situated at the macula, cause an increased hypermetropic error by pressing the retina forward. Diagnosis of choroidal malignant melanomata depends upon ophthalmoscopic examination. The retina appears dark at the region of the tumour, the shade depending upon the amount of pigment in the tumour, and it is raised above the level of the general retina The swollen retina looks round and solid, but there is usually an adjacent area of fluid

These tumours are supplied and drained by dilated and tortuous retinal vessels. Retinal angiomata are considered to be derived from congenital vascular nests and they are sometimes associated with angiomata of the cerebellum, spinal cord, pancreas, and kidney. This association is known as the Von Hippel-Lindau disease, and was included by van der Hoeve under the group of the Phakomatoses. An angioma of the retina induces the formation of glial tissue which, like other fibrous tissue, may contract causing retinal separation, and many of these eyes shrink subsequently to the state of phthisis bulbi. The retinal angiomata are treated by radiotherapy or by diathermy applications either to the tumour or to the blood vessel supplying or draining it.

2. Malignant Tumours

Retinoblastoma (Glioma of the retina; neuro-epithelioma retinæ) (Fig. 63 and Fig 64). This tumour occurs in children during the first 4 years of life and appears to be developed from imperfectly differentiated portions of the retina. Some cases show a familial incidence and about 25 per cent of cases have both eyes affected.

Pathology. The retinoblastoma is composed of small round cells showing many mitotic figures which are packed around blood vessels. The cells placed more than a certain distance from the blood vessels undergo necrosis, giving the appearance known as pseudo-rosettes. Parts of the growth show some differentiation into rosettes, which are groups of elongated cells arranged radially around a central cavity. The nuclei lie at the outer ends of the elongated cells, while the centrally directed ends become pointed near the central cavity, which is demarcated by a distinct membrane, and may contain rod shaped processes from the cells. The structure bears a resemblance to the rod and cone layer of the retina. The tumour mass may originate in the inner part of the retina when it invades the vitreous cavity and spreads seedling growths to other parts of the eyeball (Glioma endophytum), or alternatively it may commence in the outer part of the retina when the main mass of the tumour lies between the retina and choroid (Glioma exophytum). Extension of the tumour occurs along the optic nerve to invade the base of the brain, and this invasion results in death. Some advanced cases, however, may show invasion through the sclera and thence to the regional lymph glands.

Symptoms and Signs. No symptoms of this condition are commonly found, and children are brought for advice because the parents have noticed a white reflex in the pupil (the cat's eye reflex). The child in some cases develops a squint and this excites attention. The same stages of growth occur with a retinoblastoma as with a malignant melanoma of the uveal tract. The stage of symptomless growth varies in length, but the diagnosis may be made at any time by the appearance of a white reflex in the pupil or by the use of an ophthalmoscope when white lustrous plaques and, sometimes, separated retina may be seen in the fundus, mixed with the normal red reflex. The plaques, often quite small, may show new blood vessels, and they may be craggy or fluffy in appearance. The stage of raised intraocular tension causes pain. Many of these eyes have a small degree of raised intraocular tension which may cause some slight expansion of the eyeball without inducing severe pain, before the diagnosis is made. The stage of extraocular extension is not very clearly defined, since the extension initially takes place along the optic nerve and causes no sign. Extension through the sclera and cornea results in proptosis, and in an ulcerated tumour. General dissemination rarely occurs, but the regional lymph glands are affected after the sclera or cornea has been perforated, and deposits

lungs. The prognosis is worse when local extraocular extension has occurred, because a local recurrence may occur in the orbital tissues. It is characteristic of this type of tumour that after removal of the primary growth the disease may remain quiescent for many years before causing death by general dissemination. The prognosis in malignant melanomata of the uveal tract is better than in that of malignant melanomata of the skin. The reason for this is uncertain, but it has been suggested that it is due to a different derivation of the cells in the two types of tumour.

(*b*) Metastases from Carcinomata. These occur in the choroid from primary tumours of the breast, lung, stomach, thyroid gland, prostate gland, and other organs.

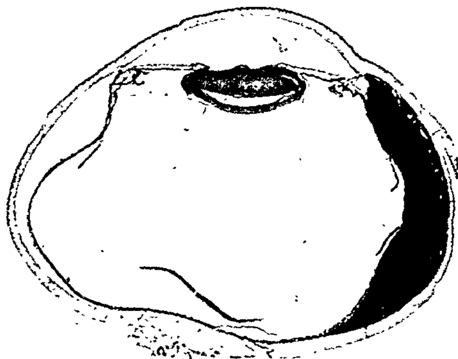


Fig. 62. Sagittal section of the eyeball showing a metastatic carcinoma of the choroid. (N. H. Ashton.)

The metastasis, not infrequently, may be the first manifestation of a previously unsuspected primary growth. It occurs usually near the posterior pole of the eye and spreads in the choroid without breaking through the membrane of Bruch. Fluid separation of the retina may develop. Metastases in the choroid appear during the stage of general dissemination and impending death, and are frequently bilateral. They lead to secondary glaucoma and radiotherapy may be useful to prevent pain and to preserve vision. These metastases are, usually, very susceptible to irradiation. Painful and blind eyes must, of course, be enucleated.

Retina

I. Benign Tumours

Angioma. Antiomata of the retina occur as small tumours, about the size of the optic disc or slightly larger, which may be multiple in an eye, and which may be bilateral.

These tumours are supplied and drained by dilated and tortuous retinal vessels. Retinal angiomata are considered to be derived from congenital vascular nests and they are sometimes associated with angiomata of the cerebellum, spinal cord, pancreas, and kidney. This association is known as the Von Hippel-Lindau disease, and was included by van der Hoeve under the group of the Phakomatoses. An angioma of the retina induces the formation of glial tissue which, like other fibrous tissue, may contract causing retinal separation, and many of these eyes shrink subsequently to the state of phthisis bulbi. The retinal angiomata are treated by radiotherapy or by diathermy applications either to the tumour or to the blood vessel supplying or draining it.

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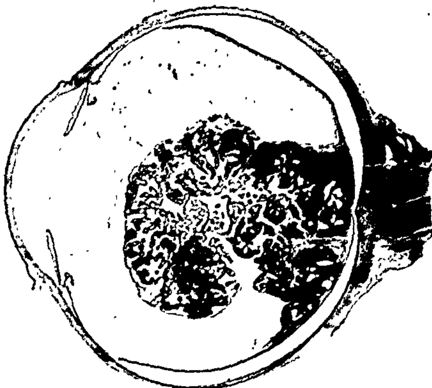


FIG 63 Sagittal section of the eyeball showing retinoblastoma. (N. H. Ashton.)



FIG. 64. Ophthalmoscopic appearance of early retinoblastoma. (H. B. Stallard.)

may be found in the bones of the cranium. Blood-borne dissemination to the lungs, bones, and other tissues has been described.

Differential Diagnosis. Retinoblastoma must be distinguished from the so-called "pseudo-glioma." This manifestation of degeneration in the eyeball follows an inflammation of the eyeball, often in utero. Formation of a cyclitic membrane leads to retinal separation, and, since it comes to lie behind the lens, to a "cat's eye reflex." These eyes are slightly shrunken, with a soft tension, and ophthalmoscopic examination shows no normal red reflex as is usually visible between the plaques of the retinoblastoma. Retinoblastoma must also be distinguished from retrolental fibroplasia (page 106). The history of prematurity and of a stay in an oxygen tent is helpful in these cases, as is also the slightly shrunken eyeball and, as a rule, the absence of any red reflex in the fundus. It is reasonable that, in cases of doubt but where the eye is considered to be sightless, the eyeball should be removed.

Treatment. Enucleation of the eyeball is required, and the optic nerve should be divided as far back as possible. Microscopic examination of the cut end of the optic nerve should be undertaken to determine if the retinoblastoma has extended to this point. Extension beyond the point of section is an indication for the orbit to be opened from above by the Naffziger method in order that the optic nerve may be removed as far back as the optic chiasma. If the chiasma has been invaded radiotherapy offers the only prospect of cure. Exenteration of the orbit may also be undertaken in patients who show an invasion beyond the sectioned end of the optic nerve, and this should also be carried out if the sclera has been invaded. Patients who have both eyes affected should have the worse enucleated and the second one treated by irradiation, by radon seeds, by radioactive tantalum wire, or by deep X-rays.

Prognosis. Cases in which the affected eyeball is removed before the optic nerve is invaded have a good prospect of recovery. The others die within 2–3 years from the effects of the growth in the base of the brain. Patients who recover from retinoblastoma should be advised against having children since there is a danger that the condition is hereditary. Parents who have had one affected child should, likewise, be advised that further siblings may also be affected.

Tumours of the Ocular Adnexa

EYELIDS

1. BENIGN. Various types of benign tumours may occur upon the skin of the eyelids as they may occur on the skin of other parts of the body. They include papilloma, xanthoma, molluscum contagiosum, angioma, and simple melanoma. The structure of these tumours is not different to their structure when they occur on other parts of the skin surface, and, with the exception of the molluscum contagiosum, treatment is not a matter of urgency and is carried out only for cosmetic reasons. Molluscum contagiosum, an umbilicated swelling divided into sections each containing large numbers of molluscum bodies, must be treated more urgently since it may spread to affect the adjacent portions of skin surface. Excision is the treatment of choice for single nodules, but for multiple nodules incision and carbolization may be a more satisfactory form of therapy.

2 MALIGNANT. (*a*) *Rodent ulcer.* This locally malignant but non-disseminating tumour is found commonly upon the skin surface of the eyelids in the middle aged and elderly. Its structure has already been described (page 110) and it appears on the eyelids

as a small nodule covered with a crust. This is shed at intervals and leaves an ulcerated surface with rolled edges which gives a serosanguinous discharge until the scab reforms. Treatment is either complete excision, which is not difficult in the early stages, or radiotherapy. The more advanced growths are difficult to excise without causing considerable



Fig. 65. Rodent ulcer of the face, in the region of the inner canthus (M Hulbert)

damage to the eyelids, while tumours at the inner end of the eyelids, though they can be treated satisfactorily by radiotherapy, often result in great obstruction to the canaliculi and in marked epiphora. The prognosis of the tumours localized to the soft tissues is good. Rodent ulcers which have advanced through the soft tissues to invade the bone or cartilage are difficult to eradicate, and the nose and its accessory sinuses may be opened to the surface.

(*b*) *Epithelioma.* This tumour usually occurs on the margin of the eyelid, in contradistinction to the rodent ulcer, which occurs on the skin surface. It may commence as a small swelling of the margin of the eyelid, but it soon ulcerates and shows a firm rolled border. The structure is the same as that of epitheliomata in any part of the body. The epithelial cells proliferate and grow through the basement membrane to invade the

dermis. This tumour is characterized by the presence of "cell nests" which show a small mass of keratin in the centre surrounded by immature epithelial cells. Some of these which are connected to each other by protoplasmic bridges are called prickle cells. Epitheliomata of the eyelid are of a low-grade malignancy, and although they may invade



FIG. 66 Microscopic appearance of mixed tumour of the lacrimal gland. (N H Ashton)

the regional glands, this is unusual. Removal of the tumour is a convenient and satisfactory treatment if it is small, but excision of the larger ones causes considerable problems of repair, and radiotherapy may be more convenient. Its results are satisfactory.

(c) *Sebaceous Carcinoma.* This tumour is derived from the sebaceous glands of the eyelid. It has, histologically, a cellular appearance, but it is not highly malignant and like the rodent ulcer it does not, in the majority of cases, lead to any general dissemination. Treatment should be by local removal or by radiotherapy.

(d) *Malignant Melanoma.* This tumour can occur in the skin of the eyelids and it may start in a pre-existing mole. The cellular structure is polyhedral or spindle in type, and irregular in arrangement, having primitive blood vessels, showing mitotic figures, and often containing melanin. Progress is usually rapid and dissemination by the blood stream occurs early. Excision is the treatment of choice, and this may be followed by radiotherapy.

Lacrimal Apparatus

MIXED CELL TUMOUR OF LACRIMAL GLAND. The only common tumour which involves the lacrimal apparatus is a mixed cell tumour of the same type as occurs in the parotid gland (which is also found in parts of the upper jaw). These tumours, which may occur in early adult or middle life, are epithelial in origin and they contain epithelial cells of varying maturity, partially arranged as glandular acini and partly in solid formation.

Myxomatous tissue is present, and there are a few areas of anaplastic tissue with many mitoses. Some capsule is present in the early stages. Growth is slow, and, at the beginning, benign in type, but recurrence may occur after partial removal, and in a more rapidly growing and malignant form. Local extension into the cranial bones and metastases both to lymph glands and by the bloodstream may occur. Adenocarcinomata of the lacrimal gland may also occur.

Tumours of the lacrimal gland appear as painless swellings protruding down behind the outer third of the upper orbital margin. They may cause downward displacement of the eyeball, defect of movement, and some degree of double vision.

Treatment necessitates complete removal of the tumour and the surrounding lacrimal gland tissue. Frequently this may mean removal of the entire lacrimal gland. The approach should be from above and requires removal of the orbital margin in order to give a full exposure of the lacrimal gland. Attempts to remove the tumour from the front are not successful, in most cases, and fail to give complete removal. Recurrence therefore is more likely. Some cases require exenteration of the orbit.

Tumours of the Orbit. Tumours of the orbit do not occur commonly. They are noticed either because they cause the eyeball to protrude or because they exert pressure on some part of the contents of the orbit. Pressure on the optic nerve may cause defective vision, while pressure on the naso-lacrimal duct may cause watering of the eye, and pressure on the fronto-nasal duct may cause acute frontal sinusitis.

Tumours of the Orbital Contents

1. Benign. (*a*) *Dermoid Cyst.* This developmental anomaly is common in the orbit in greater or lesser degree. Small ones occur at the outer and upper corner of the orbit, while larger ones may be attached deeply on the medial side. They contain a sebaceous material, and the walls show stratified squamous epithelium, hair follicles, and sebaceous cysts. They should be removed by an intracranial approach, through the roof of the orbit.

(*b*) *Angioma.* This tumour occurs fairly frequently, and it is often situated within the muscle core and may cause intermittent proptosis. It is encapsuled and can be removed fairly easily by a lateral approach.

(*c*) *Varix.* This is composed of dilated veins which become very congested with blood when the head is lowered, as in tying shoe-laces, and it causes the eyeball to protrude. The veins are usually present in the upper or lower eyelid, and they extend back into the orbit. The orbit must be opened from the front and the dilated veins excised.

(*d*) *Neurofibroma.* This occurs as part of a neurofibromatosis affecting other parts of the body, often the temporal fossa. Removal can be carried out either by the lateral or transcranial approach.

2. Malignant. (*a*) *"Mixed" Tumour of Lacrimal Gland* (page 119). (*b*) *Sarcoma.* This may occur in any of the mesoblastic tissues of the orbit and may be impure, forming a chondrosarcoma, or a fibrosarcoma, or a rhabdomyosarcoma. It progresses steadily causing proptosis and it necessitates exenteration of the orbit. Blood-borne metastases may form.

(*c*) *Glioma of the Optic Nerve.* This tumour which usually occurs in children is pathologically a fibrillary astrocytoma. It is locally malignant, but it does not cause metastases. It causes a fusiform swelling of the optic nerve which may cause enlargement

of the optic foramen. Loss of vision is an early symptom and later the eye becomes prominent without being displaced horizontally or vertically. Treatment involves removal of the affected part of the optic nerve. It may be possible to preserve a movable eyeball, though it will of course be sightless. The orbit must be approached from above and it may be necessary to remove the optic nerve back to the optic chiasma. Failure to remove the tumour may result in death from pressure on vital structures at the base of the brain, since it will progress steadily towards the optic chiasma. Some of these tumours spread to the other optic nerve by way of the chiasma. Complete removal leads to a satisfactory prognosis.

(*d*) *Endothelioma of the Optic Nerve (Meningioma).* This condition occurs in adults. These tumours are composed of oval or spindle cells, some of which may be arranged in whorls. They are locally malignant, but metastases are very rare. Axial proptosis of the eyeball is usually the first sign of the condition, and is followed by visual defect. Removal of the tumour, which rarely extends through the optic foramen, can usually be carried out by a superior transcranial approach. The eyeball may be conserved.

(*e*) *Lymphomata.* These lymphocytic tumours may occur in the orbit. They are sometimes associated with lymphatic leukæmia and they may be the early manifestation of lymphosarcoma. They consist of a mass of lymphocytes which may cause protrusion of the eyeball. They are satisfactorily treated by radiotherapy.

Tumours arising from the Wall of the Orbit and from the Nose and Nasal Sinuses

1. Benign. (*a*) *Osteoma.* This tumour may be of ivory or cancellous bone and its progress is slow. It causes proptosis and displacement of the eyeball, but diplopia is rare. It may cause obstruction of the naso-frontal duct, and it may extend to the meninges and cause meningitis and brain abscess. These tumours can be satisfactorily removed, and the approach depends upon the position of the tumour.

(*b*) *Fibroma.* This may arise in connection with the orbital periosteum and may cause some displacement of the eyeball. Removal is straightforward, through an incision made in the neighbourhood of the tumour.

(*c*) *Chondroma.* This is of the usual structure of these tumours and is attached to the orbital wall. Removal presents no difficulty.

2. Malignant. (*a*) *Osteogenic Sarcoma.* This arises in association with the orbital periosteum and is of the usual structure of sarcomata, being round celled or spindle celled. It is sometimes associated with Paget's disease of bone. It causes protrusion of the eye. The condition can be treated by exenteration of the orbit followed by radiotherapy or by radiotherapy alone. Blood-borne metastases may occur.

(*b*) *Carcinoma of Nasal Sinuses.* Carcinomata of the nasal sinuses may invade the orbit. They are usually epitheliomata of the antrum or of the ethmoidal air cells and they cause proptosis as well as the nasal symptoms. They are usually treated by fenestration of the hard palate and removal of as much of the tumour as possible followed by radiotherapy. Primary carcinomata of the frontal sinus are rare, but this structure is sometimes affected by metastases which in turn may affect the orbit.

(*c*) *Mixed Cell Tumours of the Maxilla.* These tumours, which are similar to the mixed tumours of the lacrimal and salivary glands, occur in the maxilla. Removal is undertaken by the method of fenestration of the palate and excision of the tumour, but

recurrence may take place, and may invade the orbit causing pressure on the optic nerve, and paralysis of the oculomotor muscles. It has a tendency to spread across the inferior surface of the skull, affecting the various nerves as they leave the cranial cavity.

(*d*) *Carcinoma of the Naso-pharynx.* These growths may, in a small proportion of cases, cause protrusion of the eyes and diplopia before nasopharyngeal symptoms are



FIG 67 Osteoma of the orbit anteroposterior X-ray appearance (L Ormerod)

noticed. They are usually epitheliomata and they invade the orbit through the inferior orbital fissure or through the posterior ethmoidal or sphenoidal sinuses. Proptosis is an early symptom and is usually forwards and upwards while the palpebral fissure becomes oblique running upwards and outwards. Palsy of the extrinsic ocular muscles, visual defect and neuralgia are common symptoms. Radiography establishes the diagnosis and radiotherapy is the treatment of choice.

SECONDARY TUMOURS OF THE ORBIT

1. TUMOURS EXTENDING FROM THE CRANIAL CAVITY.

(*a*) *Meningioma of Sphenoid.* This tumour may arise from the greater or lesser wing of the sphenoid and may cause proptosis by invasion of the orbital cavity or by interference with the venous or lymphatic drainage The meningioma has the usual structure

(page 121) and its progress is slow. Diagnosis is made by radiography. These tumours are not highly malignant. Partial excision may be possible, but this cannot be assisted by radiotherapy since they are not sensitive to this treatment.

(*b*) *Pituitary Tumours.* These may occasionally invade the orbit. Eosinophilic adenomata of the anterior lobe may give symptoms and signs suggestive of a sphenoidal meningioma.

2. Metastases of Tumours of other Parts of the Body.

(*a*) *Neuroblastoma* (Hutchinson's adrenal tumour). This tumour occurs in children and it arises in the adrenal or in any part of the sympathetic nervous system or from the anlage of that system. Protrusion of the eyes may be an early symptom and associated with many dilated and branching venules of the eyelids. The tumour is composed of small round cells with rosette formations. Treatment is by radiotherapy but metastases are widespread and death occurs rapidly.

(*b*) *Chloroma.* This condition occurs in the young in association with myeloid leukæmia. The cellular masses of immature leucocytes have, macroscopically, a greenish colour. Similar masses are found in many of the bones of the body and lead rapidly to death.

(*c*) *Metastatic Carcinoma.* This occurs in the orbit in the late stages of dissemination of carcinoma of the breast, of the lungs, of the gastrointestinal tract, and of other organs. Proptosis and palsy of the extrinsic ocular muscles may occur, and pressure on the optic nerve may cause a visual defect. The diagnosis is made with the knowledge that a primary growth has been present and, in some cases, that other metastases have occurred.

GLAUCOMA

Glaucoma is that condition of the eye in which the intraocular pressure is raised above what is considered to be the normal. The intraocular pressure is variable in health but it is usually between 20 and 27 mm. of mercury: a pressure above 32 mm. of mercury is regarded as abnormal. The pressure may vary by about 4 mm. of mercury during the 24 hours of the day and it is usually at its highest in the morning and at its lowest in the evening.

Measurement of the Intraocular Pressure

Accurate measurement of the intraocular pressure requires the insertion of a hollow needle or of a cannula into the anterior chamber and the attachment of this to a manometer. This is not possible as a routine in clinical practice, and two other methods are generally employed: (1) Palpation with the fingers; (2) The Schiotz tonometer.

(1) **Finger Palpation.** The patient is instructed to look down, keeping his eyes open, in order that the eyelids are relaxed and that the sclera becomes available for palpation by the fingers. The middle and fourth fingers of the observer's hands are rested on the patient's forehead and his eyeball is palpated with the two index fingers as though eliciting fluctuation. The eyeball with normal intraocular pressure gives a definite amount of fluctuation which is learnt by the experience of palpation of many eyes. The eyeball with very high pressure feels like a pebble, while that which has a low pressure has the consistency of jelly. Intermediate pressures can be recognized.

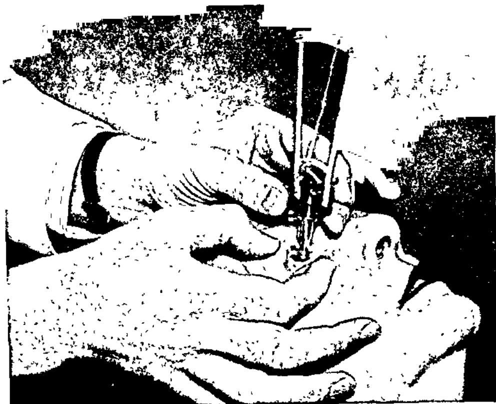


FIG 68. Estimation of intraocular pressure by the Schiotz tonometer. (K. Wybar)

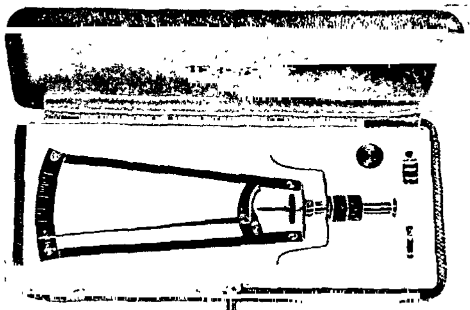


FIG 69 Tonometer (Hamblin.)

(2) The Schiotz Tonometer. This instrument is a hollow tube which has a base which is shaped to fit on the cornea. A rod in the hollow tube rests on the cornea and indents it by a varying amount according to the intraocular pressure. The amount of indentation is indicated upon a gauge and the intraocular pressure can be calculated on a graph, which has been prepared by observing the indentation of the cornea caused by known



FIG. 70. Glaucomatous cupping of the optic disc. (Hansell)

weights on eyes in which the intraocular pressure is known from a cannula and manometer reading. Readings of the intraocular pressure registered by the Schiotz tonometer may not be absolutely accurate, but they give an approximate determination, and the pressures taken by the same individual with the same tonometer are valuable for purposes of comparison. The use of the Schiotz tonometer necessitates that the cornea is anæsthetized with one or two drops of 1 per cent amethocaine. Cocaine hydrochloride drops should not be used because they dilate the pupil and this may cause a rise of the intraocular pressure. The patient is made to lie on a couch, so that the eye looks directly upwards, and the tonometer is placed vertically upon the cornea and the reading is observed. Care must be taken not to cause a corneal abrasion. An oily drop, such as castor oil or paroleine, should be inserted after the reading has been made.

Classification

(1) Primary Glaucoma. The condition of raised intraocular pressure without any causative ocular disease. There are two types, which are distinguished, essentially,

according to the type of angle of the anterior chamber, as observed by gonioscopy examination. This examination is carried out with a special contact lens which is placed in the patient's eye, after which examination with the slit lamp and corneal microscope allows accurate observation of the angle:

(*a*) Acute congestive glaucoma (narrow angle glaucoma), characterized by the early onset of high intraocular pressure, of corneal œdema, of congestion and of pain. Cupping of the optic disc may be present in advanced cases.

(*b*) Chronic simple glaucoma (wide angle glaucoma), in which the intraocular pressure is raised by a smaller amount, than in the congestive form, in which there is no pain or congestion, and in which cupping of the optic disc is often present with contraction of the visual field.

(2) **Secondary Glaucoma.** Raised intraocular pressure associated with and caused by some other ocular disease.

(3) **Infantile Glaucoma.** Raised intraocular pressure occurring in infancy, which causes distension of the eyeball due to the great plasticity of the tissues of the young. The rise of pressure may be primary or secondary.

Primary Glaucoma. The cause of primary glaucoma is unknown but it seems to be associated with defective drainage of aqueous from the eye.

(*a*) Acute Congestive Glaucoma

A painful condition, of sudden onset, which usually causes a marked defect of the visual acuity. The clinical symptoms and signs fall conveniently into three stages: (1) The prodromal stage; (2) The acute congestive stage; (3) The stage of absolute glaucoma.

(1) The Prodromal Stage. This stage occurs in most cases, but the symptoms vary in severity and frequently the patients do not complain at this time, though the history may be elicited when they come under treatment at a later date. This prodromal stage is characterized by transient attacks in which the vision of the eye is blurred and in which there is discomfort in the eye with slight headache. Lights observed during this stage are surrounded by coloured rings which are known as haloes and which are due to the corneal œdema. Haloes may also be the result of small patches of mucus on the cornea and of corneal scarring, but if a patient complains of this symptom, it is essential that he should be investigated to exclude incipient glaucoma. Examination during the prodromal attacks shows faint blurring of the cornea and a semidilated pupil which may be fixed, or which may be sluggish in reaction. These attacks are transient and they usually pass in a few minutes or a few hours without treatment. They may occur in association with certain definite events such as visits to the cinema, when sitting in the dark causes the pupils to dilate and provokes the attack. They may occur also in association with emotional crises, such as worries about business, family troubles, and even after large meals or excessive drinking. They may recur at irregular intervals for a period of many years without progressing to the second stage of narrow angle glaucoma. Some patients who are having treatment with miotics may have occasional prodromal attacks, but these pass quickly if a drop of the miotic is inserted in the eye, and such patients may be kept on miotic therapy for many years without damage occurring to the optic nerve.

(2) The Acute Congestive Stage. This stage may occur acutely, either as a sequel to a prodromal attack or without such attack, and it is frequently associated with some serious emotional disturbance. Ocular pain is severe in most cases and the visual acuity

is much reduced. The pain may spread from the eye to the forehead and to the upper jaw, and it may be so severe as to cause constitutional symptoms such as nausea, vomiting, abdominal pain, and collapse. Cases are recorded in which patients in the acute congestive stage of narrow angle glaucoma have been diagnosed as suffering from some acute abdominal emergency. Examination of the eye reveals a rise of intraocular pressure, the eyeball having a stony feeling on palpation. The conjunctiva is congested and some chemosis may be present while the cornea is hazy, due to the œdema of the epithelial cells. The cornea may be insensitive. The anterior chamber is usually shallow and the pupil is semi-dilated and fixed. The iris is congested and œdematous. Fundus examination with the ophthalmoscope is not possible on account of the opacity caused by the corneal œdema. These eyes may suffer complete loss of vision within a few days if not treated. Occasionally, in the less severe cases, the loss of vision may be more gradual, and, sometimes, a remission takes place so that the intraocular pressure returns to normal, though some damage to the eyeball has usually occurred. Subsequent attacks may take place which cause more and more damage to the eyeball. Prolonged rise of intraocular pressure causes cupping of the optic disc. The lamina cribrosa is that part of the sclera which is present at the site of entry of the optic nerve, and it is the weakest section of the uninjured eyeball. Continued rise of intraocular pressure causes it to bulge backwards with the fibres of the optic nerve, and this may cause injury to some of these fibres. This bulging of the lamina cribrosa can be seen by the ophthalmoscope as a deep pale excavation of the optic disc. The central vessels of the retina may appear in the middle of the cup running on its floor and they can also be focused at a higher level on the rest of the retina. Pulsation of the arteries is a characteristic sign of raised intraocular pressure.

(3) The Stage of Absolute Glaucoma. Prolonged rise of intraocular pressure leads to blindness and absolute glaucoma. The congestion of the eyeball disappears, the cornea clears, and the pupil is widely dilated and immobile. The anterior chamber is shallow and the intraocular pressure remains high. The optic disc is excavated and pale, and it is surrounded by the white glaucomatous ring, which is due to atrophy of the retinal and choroidal pigment around the disc. Pain may disappear, but it sometimes remains as a constant recurrent symptom.

Treatment

Prodromal Stage. Patients seen at this stage should be treated with miotic drops which contract the pupil and open up the angle of the anterior chamber. Pilocarpine nitrate drops 1 per cent or 2 per cent or eserine salicylate drops ¼ per cent, ½ per cent, or 1 per cent should be used two or three times daily. These drugs can be combined if circumstances require it. Sedative treatment with phenobarbitone gr. ½ twice daily may be useful. Surgical treatment should be undertaken if the prodromal stage persists in spite of treatment with miotics, a peripheral iridectomy being all that is required in most patients.

Congestive Stage. Patients in this stage of glaucoma should be treated in hospital, or, if it is necessary to treat them at home, it is essential to provide *full nursing facilities*. The treatment is: (1) Medical; (2) Surgical.

(1) *Medical Treatment.* The patient is placed at rest in bed and he is treated with miotic drugs, eserine salicylate drops 1 per cent usually being necessary. The drops are inserted in the affected eye every 5 minutes for an hour, every hour for 4 hours, and then

4 hourly, while the unaffected eye receives a drop every 4 hours to prevent the development of congestive symptoms. Applications of heat by hot bathing or by short-wave diathermy are helpful, and analgesic drugs such as omnopon gr. 1/3 are usually required to overcome pain. Diamox (Acetazolamide) is a drug of the greatest value in these patients. It has the effect of reducing the flow of aqueous into the eye, if given by mouth in doses of 250–1,000 mg. daily, and it causes a reduction of the intraocular pressure. The eye should be re-examined after 12 hours of treatment. A normal intraocular pressure is a sign that medical treatment has been successful, and that this should be maintained until the eye becomes settled, when the possibility of surgical treatment to prevent further

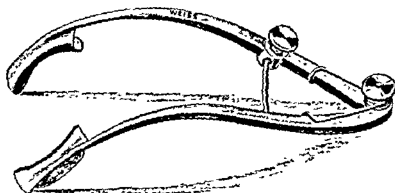


Fig. 71. Eye speculum.

attacks of congestive glaucoma must be considered (page 130). An eye with a persisting high intraocular pressure after 12 hours of treatment requires urgent surgical treatment.

(2) *Surgical Treatment.* The most satisfactory operation for acute congestive glaucoma is the classical iridectomy of von Graefe. *Instruments*: Eye speculum (Fig. 71), fixation forceps, narrow cataract knife, iris forceps, iris scissors (Fig. 51), and iris repositor. *Anæsthetic*: A general anæsthetic is desirable when possible, but the operation can be performed under the influence of 4 per cent cocaine hydrochloride drops and with 1 c.c. of 4 per cent procaine injected behind the eyeball into the cone of muscles. 2 c.c. of 4 per cent procaine should be injected into the region where the branches of the facial nerve cross the neck of the mandible because this causes paresis of the orbicularis oculi muscle. *Operation*: The surgeon stands (or sits) behind the patient's head as he lies on the operating table and introduces the speculum. The internal rectus muscle is grasped with fixation forceps and the eye is held so that it is directed downwards. It is necessary that tension is kept upon the fixation forceps so that they do not press on the eyeball. The point of the cataract knife enters about 1 mm. behind the limbus at the junction of the upper 1/6 and lower 5/6 of the cornea, and it is passed across the anterior chamber to come out at a corresponding point on the opposite side. Then the section is completed. Iris forceps are passed into the anterior chamber and the iris is grasped. It is cut through at one end of the wound and the cut end is drawn out in such a manner that the iris is torn away from its attachment to the ciliary body, until the other end of the section is reached, when it is again cut through, and the separated portion removed. The pillars of the remaining iris must be replaced in the anterior chamber and free of the margins of the wound. Atropine is instilled to the operated eye and eserine to the other eye. Both eyes are bandaged, and kept so for 24 hours. The unoperated eye may be uncovered at

the end of this period, but it must continue to receive daily drops of eserine. The operated eye must be dressed and a drop of 1 per cent atropine sulphate inserted each day until the wound is well healed.

Complications: (1) *Post operative uveitis.* This follows all intraocular operations, but it is only serious when it is severe in degree, and when it persists for many days and even weeks after the operation. It is more persistent if one of the iris pillars is left adherent to the wound, because this causes continued irritation. (2) *Expulsive hæmorrhage.* This very profuse hæmorrhage occurs inside the eye as the corneal section is completed. The sudden lowering of intraocular pressure may be a causative factor, and the section should, therefore, be made very slowly. (3) *Traumatic cataract.* This is caused by rupture of the anterior lens capsule either by the point of the cataract knife, by the iris forceps, or by the repositor. Removal of the cataract with recovery of vision is possible.

(*b*) Chronic Simple Glaucoma

This condition differs from acute congestive glaucoma in being insidious and painless in its onset and in its course, and in causing little or no impairment of visual acuity in the early stages. The diagnosis is usually made at a fairly advanced stage when the patient notices some visual defect and attends for routine testing of the refractive error. The

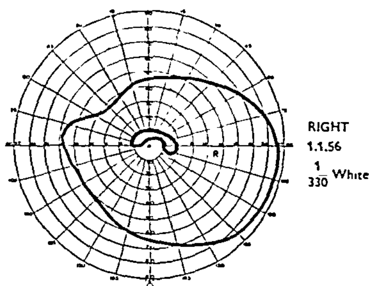


Fig 72. Visual field chart of a patient with open angle glaucoma showing an arcuate scotoma and some constriction of the upper nasal field

ophthalmoscopic examination reveals a pale cupped disc, and the intraocular pressure may be found to be raised. The important diagnostic point in this disease is the loss of a portion of the visual field, which is estimated on the perimeter (Fig. 57) and on the Bjerrum screen (Fig. 58). The average field of vision extends 60 degrees on the nasal side, 55–60 degrees above, 75 degrees below, and 100 degrees temporally, and it contains the blind spot which corresponds to the optic disc 15 degrees temporally from the point of fixation. The most common forms of visual field defect which occur in chronic simple glaucoma are some loss of the upper peripheral nasal field, and the formation of the

4 hourly, while the unaffected eye receives a drop every 4 hours to prevent the development of congestive symptoms. Applications of heat by hot bathing or by short-wave diathermy are helpful, and analgesic drugs such as omnopon gr. 1/3 are usually required to overcome pain. Diamox (Acetazolamide) is a drug of the greatest value in these patients. It has the effect of reducing the flow of aqueous into the eye, if given by mouth in doses of 250–1,000 mg. daily, and it causes a reduction of the intraocular pressure. The eye should be re-examined after 12 hours of treatment. A normal intraocular pressure is a sign that medical treatment has been successful, and that this should be maintained until the eye becomes settled, when the possibility of surgical treatment to prevent further

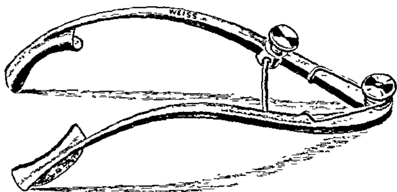


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(*b*) Chronic Simple Glaucoma

This condition differs from acute congestive glaucoma in being insidious and painless in its onset and in its course, and in causing little or no impairment of visual acuity in the early stages. The diagnosis is usually made at a fairly advanced stage when the patient notices some visual defect and attends for routine testing of the refractive error. The

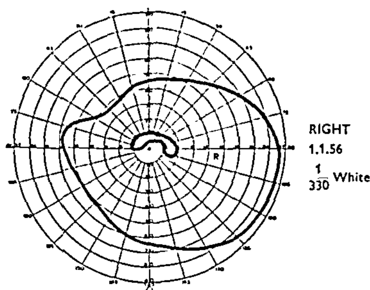


Fig. 72. Visual field chart of a patient with open angle glaucoma showing an arcuate scotoma and some constriction of the upper nasal field.

ophthalmoscopic examination reveals a pale cupped disc, and the intraocular pressure may be found to be raised. The important diagnostic point in this disease is the loss of a portion of the visual field, which is estimated on the perimeter (Fig. 57) and on the Bjerrum screen (Fig. 58). The average field of vision extends 60 degrees on the nasal side, 55–60 degrees above, 75 degrees below, and 100 degrees temporally, and it contains the blind spot which corresponds to the optic disc 15 degrees temporally from the point of fixation. The most common forms of visual field defect which occur in chronic simple glaucoma are some loss of the upper peripheral nasal field, and the formation of the

so-called arcuate scotoma passing from the blind spot above or below the point of eye fixation (Fig. 72). These defects are due to lesions of the fibre bundles of the optic nerve at the extreme anterior end of the optic nerve, though whether their atrophy is directly due to pressure or to a defect of the vascular supply is uncertain.

Chronic simple glaucoma may be difficult to diagnose in its early stages, and certain tests are helpful:

(1) PHASING. The estimation by the Schiotz tonometer of the intraocular pressure every 2 hours for a period of 24 hours. A rise of the intraocular pressure above 32 mm. of mercury is suggestive of glaucoma, as is a variation of more than 4 mm. of mercury in the 24 hours. The phasing can be repeated while the patient is being treated with miotic drops to estimate their effect.

(2) ESTIMATION OF AQUEOUS OUTFLOW. This can be carried out by various methods. The principle is that if pressure is applied to an eyeball the intraocular pressure falls due to drainage of aqueous. A low outflow of aqueous, as shown by only a small reduction of intraocular pressure, indicates that aqueous drainage is defective and, even if the intraocular pressure is normal, that danger of a rise of the pressure exists and that surgical treatment may be required.

(3) PROVOCATIVE TEST. The drinking of a litre of water with measurement of the intraocular pressure before and after has been employed to assess the presence of glaucoma.

TREATMENT. (1) *Medical Treatment.* This is carried out with miotic drops such as ¼ per cent, ½ per cent, or 1 per cent eserine salicylate drops, or 1 per cent or 2 per cent pilocarpine nitrate drops. The effect of the drops can be estimated most satisfactorily by phasing the patient while the drops are being used, but it can also be assessed by ordering the drops for regular use and by observing the patient at regular intervals for abnormal increase of intraocular pressure and for progressive loss of visual field. Uncontrolled intraocular pressure and progressive loss of visual field are indications for surgical treatment, and a very defective aqueous outflow may also be an indication for operation. The use of sedation is helpful in these patients, and phenobarbitone ½–1 gr. twice daily may help to control the rise of intraocular pressure. Diamox has little place in the treatment of wide angle glaucoma unless there is an acute exacerbation of pressure.

(2) *Surgical Treatment.* Various operations are used in the treatment of wide angle glaucoma. The classical iridectomy is valueless because in this type of glaucoma the iris does not cause obstruction to the drainage of the aqueous. The operations most commonly used are (1) corneoscleral trephining; (2) iridencleisis; (3) anterior sclerectomy. The former of these is that which is most generally employed.

Corneo-scleral Trephining. *Anæsthetic.* This operation can be carried out under a general anæsthetic, but it is most usually performed under the local application of 1 per cent cocaine hydrochloride drops, and with an injection of 1 c.c. of 4 per cent procaine into Tenon's capsule (Retro-ocular injections reduce pressure and add to the difficulties of this operation.) *Instruments*: Speculum (Fig. 71), fixation forceps, Lister forceps, squint scissors (Fig. 73), Tooke's knife, 1·5 mm. trephine, disc forceps, disc scissors, iris repositor, needle holder, 60 silk on small curved needles *Operation*: The patient lies on the operating table, with the surgeon standing (or sitting) at the head. The speculum is inserted and a small flap of conjunctiva is raised at the upper part of the eye

and dissected to the corneal margin. The incision is carried into the cornea by splitting it with the Tooke's knife. The trephine is applied with its centre at the corneo-scleral junction and the disc is cut. The pupil is displaced upwards as the trephine enters the angle of the anterior chamber. The disc is grasped with the disc forceps and is cut off. The iris is held with the iris forceps and a peripheral iridectomy is performed, after which the

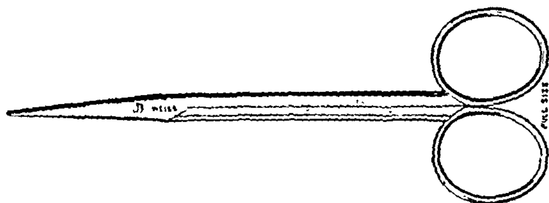


FIG. 73 Squint scissors

iris retracts into the anterior chamber. The flap is smoothed back and sutured in position. A drop of 1 per cent atropine sulphate is placed in the eye, and ½ per cent eserine salicylate is placed in the unoperated eye. A bandage is applied to the operated eye. *Complications:* (1) *Delayed reformation of the anterior chamber.* It is essential to be sure that no leak exists through the conjunctiva. A drop of fluorescein is placed in the eye, and if it appears green near the site of the trephining a leak exists. An extra conjunctival flap brought down from above is useful in cases of delayed reformation of the anterior chamber, whether a leak can be demonstrated or not. (2) *Late infection of the bleb.* A trephine bleb is liable to infection which appears to invade it from the conjunctival sac. This can be treated by the administration of systemic penicillin or of other antibiotics.

Secondary Glaucoma. The most common causes of this condition are: Anterior synechiæ following perforating injuries of the eyeball, dislocation of the lens, traumatic cataract with swelling of the lens, intraocular hæmorrhage, acute uveitis, the posterior synechiæ, seclusio pupilli, and occlusio pupilli which may result from uveitis, intraocular tumours, and obstruction of the central vein of the retina (thrombotic glaucoma).

Symptoms and Signs. These may be obscured by the primary condition, but they resemble narrow angle glaucoma, except that pain is not such a marked symptom. The intraocular pressure is raised, corneal œdema and conjunctival congestion are present, and the pupil may be dilated. The cause of glaucoma in these cases is usually obvious. The obstruction of the angle of the anterior chamber by hæmorrhage and by inflammatory exudates, and the inability of the aqueous to reach the angle in occlusio and seclusio pupilli are obvious causes. Patients who have had obstruction of the central vein of the retina develop vascular granulation tissue which spreads over the iris into the angle, which it obstructs. The cause of secondary glaucoma in patients with subluxated lenses and with intraocular tumours is not so easy to explain

Treatment. The necessary treatment of secondary glaucoma is that of the cause. Anterior synechiæ should be divided, seclusio and occlusio pupilli treated by iridotomy or by iridectomy, traumatic cataract should be washed out, and dislocated lenses may be

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appears in the non-injured (*sympathizing*) eye. It is unusual for sympathetic ophthalmitis to appear before the tenth day after the injury and, therefore, provided an injured eye is removed within this period it is unlikely that sympathetic ophthalmitis will occur. The decision regarding removal of the eye may be difficult, and borderline cases can only be decided by the experience of the surgeon. The removal of the injured eye when sympathetic ophthalmitis has already developed may not affect the progress of the condition. It should be removed, however, if its vision is absent or is very defective.

(3) **Irremovable Intraocular Foreign Bodies.** Eyes in which there is an intraocular foreign body, which cannot be removed without inflicting irreparable damage, should be removed if irritation is present. Such an eye may induce sympathetic ophthalmitis.

(4) **Panophthalmitis** (page 105). Panophthalmitis is a suppurative inflammation of the whole eye, involving the vitreous cavity. It becomes manifest by the presence of pus in the anterior chamber and in the vitreous body in an inflamed congested eye. The main indication for removal of the eye is loss of the ability to project light (to identify the direction from which light is being shone at the eye), this state arising when the retina has been detached by the pull of the cyclitic membrane. Antibiotics can help many cases of early panophthalmitis, and can save many eyes which would have required removal in former times, but once the ability to project light has been lost, no antibiotic can restore the vision of the eye.

(5) **Malignant Tumours.** The presence of a malignant tumour in an eyeball requires its removal. It is difficult in some patients to decide whether or not a malignant tumour is present, and, since the eye may possess full visual acuity, the decision to remove the eye is one of great seriousness. In doubtful cases it is justifiable to keep the patient under observation to examine possible progress since it is known that, with many malignant neoplasms of the eye, general dissemination occurs very early, and that the ultimate prognosis is not affected by a few months' delay in enucleation of the eyeball.

(6) **Blind Painful Eye.** This type of eye may have become blind following injury, severe inflammation, neoplasm, glaucoma, or other causes. Pain is due, in the majority of cases, to uveitis which may be primary, or which may be that type which often occurs in a degenerate eye. Pain may also be due to glaucoma. The continuous pain may prevent sleep and cause a general deterioration in the health of the patient, and removal of the eyeball must be strongly recommended. A considerable proportion of these eyes when examined after removal are found to contain malignant neoplasms (chiefly malignant melanomata) and this is an additional reason for recommending removal of the eyeball. A painful or irritable eye which is certain to become blind should also be removed.

(7) **Disfiguring Eyes.** A scarred or shrunken eye may be a considerable disfigurement even if it is not painful or irritable. The majority of such patients will retain such an eye and disguise its appearance with a frosted glass or an eyeshade. A minority, however, will prefer removal so that a suitable prosthesis can be fitted which can be difficult to distinguish from a normal eye.

Operations for Removal of the Eyeball

(1) **Enucleation of the Eyeball.** *Anæsthetic.* This operation should be performed under a general anæsthetic, but in an emergency it can be performed under local anæsthesia. 2 ml. of 2 per cent procaine are injected retro-ocularly into the cone formed by the rectus muscles. This may cause considerable protrusion of the eyeball, but this is not significant.

removed with the scoop or vectis. Diagnosis is of supreme importance. The identification of the glaucoma as secondary is the first necessity, but the elucidation of the cause is equally essential. Diamox given in doses of 250–1,000 mgm. daily is of great use in some cases of secondary glaucoma, especially in those due to an inflammatory cause. Paracentesis may be helpful. A broad needle is inserted into the anterior chamber and the aqueous is drained off. This may carry the patient along until the cause has settled. Cyclodiathermy is also useful in some cases. This procedure involves the application of a diathermy electrode over the ciliary body. This decreases the function of the ciliary body and lowers the intraocular pressure. The electrode is applied direct to the sclera 6 mm. behind the limbus with a current of 70 milliamperes. Applications lasting 6 seconds are made fairly closely around half the circumference of the limbus. The immediate results are satisfactory but the rise of tension may recur. The procedure can, however, be repeated over the other half of the circumference and even over the zone previously treated.

Infantile Glaucoma. Glaucoma occurring in children causes stretching of the soft pliable tissues of the child's eye, and, as a result the eyeball becomes distended. This condition is called buphthalmos. Infantile glaucoma may be a primary developmental defect, probably at the angle of the anterior chamber, or it may be secondary due to any of the causes of secondary glaucoma already enumerated. The treatment is usually surgical and a trephining or other drainage operation may be required. The operation of goniotomy, the opening of the angle of the anterior chamber by a special knife, has given satisfactory results in this condition.

REMOVAL OF THE EYE

Knowledge increases and ophthalmology from being a small branch of general surgery has progressed to the status of a specialty with its own techniques of investigation and treatment. One procedure, however, which should be within the capabilities of any registered medical practitioner is enucleation of the eyeball. This may be required as an emergency for a variety of reasons, and it is necessary that the removal should be carried out efficiently, and without leaving in the orbit any uveal tissue which might induce sympathetic ophthalmitis. It is important also that an adequate socket should be left for the fitting of a prosthesis. Simple enucleation is the operation of choice for those who do not have special experience in ophthalmic surgery, because, though many modifications of the simple enucleation, involving the attachment of implants to the extraocular muscles to give better movement, are available, their results are uncertain and they are not to be recommended without special indications.

Indications for Removal of the Eye

(1) **Severely Injured Eye** (page 89). An eye which has been injured so severely that there is no possibility of vision being restored, should be removed. It must be appreciated that badly damaged eyes can recover in remarkable fashion if they are carefully repaired, and where any doubt exists the removal should be delayed. Nevertheless in some cases recovery is manifestly impossible and removal should be carried out at once.

(2) **Prevention and Treatment of Sympathetic Ophthalmitis** (page 89). Sympathetic ophthalmitis is a form of uveitis which occurs after injury of one eye, in the other eye of a patient. It can be prevented by removal of the injured (exciting) eye before the condition

eye, usually made of plastic, is a very satisfactory prosthesis, and it can be fitted so that it does not tend to sink back into the orbit more than its fellow. It does not move as much as the normal eye, though some movement is present, but these patients can be taught to restrict their ocular movements so that the lack of mobility is not obvious.

Evisceration of the Eyeball

This operation is employed when an eyeball is very badly injured, and when severe intraocular inflammation is present. It requires a general anæsthetic. *Instruments.* Eye speculum, fixation forceps, strabismus scissors, curved if possible, small knife, and evisceration curette. *Operation.* The speculum is inserted and the knife is passed into the eyeball at the corneo-scleral junction, and passed round the cornea so as to remove it. The contents of the eyeball are removed with the evisceration curette, care being taken that no uveal tissue is left behind. No suture is inserted. A firm pad and bandage are applied. Some surgeons, after removal of the contents of the eyeball, separate the sclera and conjunctiva and remove a collar of sclera 8–10 mm. deep. This hastens healing, which is likely to be more prolonged than after enucleation, though it is to be expected that a satisfactory socket will result.

After Results of Removal of the Eye

The prosthesis should be fitted as soon as possible, and it should always be worn during the waking hours. Failure to do this results in inturning of the eyelashes which cause irritation of the conjunctiva, and excessive mucopurulent discharge. Some slight discharge occurs in all cases, but if the socket is irrigated, morning and evening, before the prosthesis is inserted and after it is removed, this causes little inconvenience. Glass prostheses are affected by the tear secretion and, after periods of about a year of constant use, become slightly roughened and this may cause discomfort and increased discharge. Polishing of the prosthesis or the provision of a new one is then indicated. Plastic prostheses last much longer but, eventually, they require renewal also. Patients, rarely, are sensitive to plastic materials. The conjunctiva of the socket becomes chemotic and the eyelids red and œdematous. Such patients must be fitted with glass prostheses.

The eyeball, in some cases, may have to be removed early in life. These small patients must be fitted with a prosthesis and this must be worn during all waking hours. The absence of the eyeball causes defective development of the bony orbit in some of these cases, with resulting facial asymmetry. Shrinkage of the socket occurs sometimes, especially if the prosthesis is not worn regularly, so that it becomes impossible for the artificial eye to be retained in the socket. Such cases must be treated by enlarging the socket and grafting the raw area with a Thiersch skin graft. It is necessary to plan the new socket very much larger than the required size, because much contracture occurs after the operation.

OCULAR MANIFESTATIONS OF GENERAL DISEASES

The eye may be affected, secondarily, by pathological processes occurring in other parts of the body. The optic nerve and the retina are, embryologically, an outgrowth of the brain and it is possible by ophthalmoscopic observation of the retina to diagnose various pathological processes occurring within the nervous system. The commonest example of this is the condition of papillœdema which occurs in association with a rise of

Instruments. Eye speculum (Fig. 71), fixation forceps, strabismus scissors, squint hook, enucleation scissors, needle holder, small curved needle, and thin black silk. *Operation.* The speculum is inserted. The conjunctiva is divided close to its junction with the cornea and it is reflected back to the insertions of the rectus muscles. Tenon's capsule is opened between the attachments of the muscles. The squint hook is placed around the attachments to the rectus muscles and they are divided, in turn. It is sometimes suggested that the external rectus muscle should be cut a few mm. from its insertion in order that it may provide a grip, but this is unimportant. The eyeball is then dislocated forwards by pressing the speculum backwards. The enucleation scissors are passed between the conjunctiva and the sclera and the optic nerve can be felt. This is divided as far back as possible. The eyeball is grasped by the fingers of the left hand, and the oblique muscles are divided to separate the eyeball from the body. A swab is immediately pushed into the socket, and pressure is applied for 1 or 2 minutes to control hæmorrhage. The conjunctival margins are brought together and are sutured vertically. This tends to give maximum size to the fornices which is helpful in the fitting of a prosthesis. A firm pad and bandage should be applied and the patient kept in bed for 2 days. The suture can be removed on the fifth day. The artificial eye can be fitted as soon as the socket is free from the bruising and the inflammation which is usually about a month after the operation.

Complications

(1) Infection

Infection occurring in the socket may spread up the optic nerve and cause meningitis. Enucleation of an eye with advanced panophthalmitis may be followed by ascending infection and meningitis, and in these cases the operation of evisceration is advised (page 135).

(2) Rupture of the Eyeball

This occurs sometimes with a friable eyeball and it makes the complete removal more difficult. It is essential that all uveal tissue should be removed, since if any is left in the orbit it is a potential cause of sympathetic ophthalmitis. Rupture of the eyeball in the presence of an intraocular tumour presents problems which are all too obvious. The primary intraocular tumour is, as a rule, circumscribed within the sclera and (though general dissemination may have occurred) it can be completely removed in the eyeball. Rupture of the eyeball during removal renders local recurrence more likely.

(3) Removal of the Wrong Eyeball

It may seem that this is an obvious mistake and that precaution would, automatically, be taken to prevent it. Nevertheless, the error is not unknown. In the case of a shrunken or scarred eye, no mistake will arise, but when the enucleation is on account of, for instance, a choroidal neoplasm, the external appearance of the two eyes is similar, and observation with the ophthalmoscope immediately before the removal is carried out is the only effective safeguard.

Orbital Implants. More effective movement and a more satisfactory appearance of the prosthesis can be obtained if an implant is placed within the cone of the rectus muscles, and if these muscles and Tenon's capsule are sewn over it. This implant may be a gold or glass sphere of the size of a large pea, or a small piece of costal cartilage, while in recent times more complicated appliances have been described. The modern artificial

eye, usually made of plastic, is a very satisfactory prosthesis, and it can be fitted so that it does not tend to sink back into the orbit more than its fellow. It does not move as much as the normal eye, though some movement is present, but these patients can be taught to restrict their ocular movements so that the lack of mobility is not obvious.

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intracranial pressure, but optic atrophy occurring in tabes dorsalis, or as a result of pressure from cerebral tumours, and choroidal tubercles occurring in tuberculous meningitis, are other diseases of the nervous system, and its covering, which manifest themselves in the fundus of the eye. Metastases of malignant tumours may be found in the eyeball and in the orbit, and defects of the visual field may indicate their presence in the various parts of the visual tract. The presence of metastases in these various situations is usually an indication of the wide dissemination of a primary and already recognized tumour, but occasionally the presence of a metastasis in the choroid may be the first manifestation of a malignant tumour, particularly of the bronchus or prostate. Certain general diseases such as hypertension and diabetes mellitus produce striking signs in the retina. The retinal appearances of hypertension are useful as an indication of the stage of severity which has been reached by the hypertension and in suggesting the onset of that advanced and serious condition known as malignant hypertension. Severe anæmia may cause fundus changes, and profuse hæmorrhage may lead to serious optic atrophy. Leukæmia is a condition which may first become apparent due to deposits of leukæmic cells in the conjunctiva or in the orbit, and it can give a characteristic appearance in the retina. Abnormalities of the endocrine glands may cause various ocular changes, and one of the most important is that prominence of the eyes, called proptosis or exophthalmos, which occurs in association with dysfunction of the thyroid gland.

Rise of the Intracranial Pressure: Papillœdema

Papillœdema, or plerocephalic œdema, is that condition of œdema of the optic nerve head which is the result of a rise of the intracranial pressure. It is postulated that in these cases cerebrospinal fluid is forced into the subarachnoid space of the optic nerve and that its pressure is sufficiently high to compress the central vein of the retina but not the central artery of the retina where it crosses this space, passing from the optic nerve to the orbit, and to cause vascular engorgement of the optic nerve head. The rise of intracranial pressure is usually due to an intracranical neoplasm, but intracranial hæmorrhage, cerebral abscess, and intracranial thrombophlebitis may also be responsible. Papillœdema may occur as a result of hypertension.

Symptoms and Signs. This condition causes few symptoms, although the underlying lesion may cause pain and defects of vision. The visual acuity may be affected by papillœdema in its later stages. The first sign of papillœdema is some blurring of the margins of the optic disc, but this progresses to swelling of the disc which is usually bilateral and which is fairly localized to the optic nerve head, the surrounding retina showing little œdema until late in the development of the condition. The veins are swollen and engorged, and there are many hæmorrhages. The later stages are accompanied by spreading retinal œdema and a star figure may be present at the macula. Secondary atrophy of the optic nerve occurs if the intracranial pressure is not relieved, and the sight may be lost.

Treatment. This depends upon the cause which must be treated without delay.

Lesions of the Visual Pathways

Recognition of the site of abnormalities of the visual pathway leads to accurate localization of intracranial lesions. This recognition depends upon accurate estimation of the fields of vision by the perimeter (Fig. 57) and Bjerrum's screen (Fig. 58), and upon

the application of a knowledge of the structure of the visual pathway. The visual impulses pass from the retina to the occipital cortex and their pathway consists of the optic nerve, the optic chiasma, the optic tract, and the optic radiation (Fig. 75). Certain visual functions depend upon connections between different parts of the cerebral cortex.

(1) Retino-occipital Pathway

(*a*) The Retina

Lesions of the retina which can often be observed by the ophthalmoscope, and may cause a defect of one visual field, are usually irregular in type. No serious embarrassment

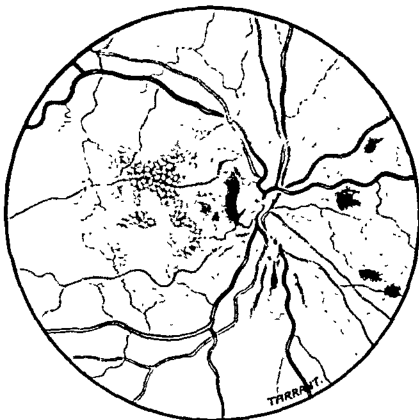


Fig 74. Ophthalmoscopic appearance of retina and optic disc in malignant hypertension. (Hansell)

of vision arises unless a very large portion of the retina is damaged, so that the peripheral field is much constricted, or unless the function of the macula is lost, when accurate central vision is obliterated.

(*b*) The Optic Nerve

A lesion of the optic nerve causes a defect of one visual field, which may be complete or incomplete, and which is usually irregular in character. It must be remembered that the position of the nerve fibres in the optic nerve corresponds, approximately, to the four quarters of the retina, so that fibres running, for instance, from the upper nasal quadrant of the retina, lie in the upper nasal part of the optic nerve. This arrangement is modified by the position of the macular fibres which enter the nerve temporally and come to lie

centrally. The nasal fibres of the optic nerve remain separate from the temporal fibres in preparation for their crossing at the optic chiasma. The lower nasal fibres loop forward into the opposite optic nerve before they pass backwards in the chiasma to the optic tract and, therefore, a lesion at the posterior part of the optic nerve may cause a complete loss of the visual field of that eye and some defect of the upper temporal field of the other

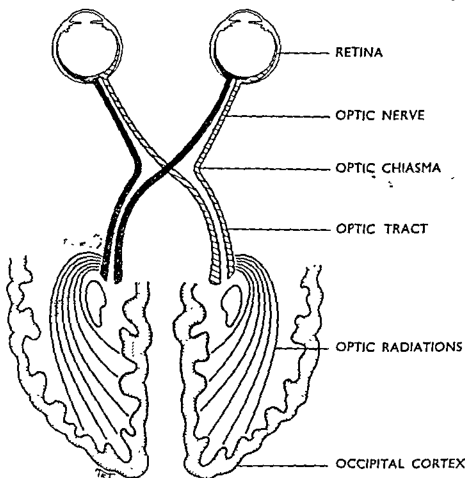


FIG. 75 Diagrammatic representation of the path of the visual impulses from the retina to the occipital cortex

eye. Lesions of an optic nerve lead to optic atrophy, manifest by pallor of the optic disc, which can be observed by the ophthalmoscope.

(c) THE OPTIC CHIASMA

The optic chiasma is situated on the upper surface of the body of the sphenoid in a position which varies slightly in different persons. The nerve fibres from the nasal portions of both retinæ cross in this structure and pass into the optic tract of the opposite side where each joins the uncrossed temporal fibres of the other eye. Lesions of the optic chiasma, unless very small, cause defects in both fields of vision, the most characteristic defect being the bitemporal field loss, which may become a complete bitemporal hemianopia (Fig. 77), and is the result of pressure upon, and atrophy of the crossing nasal fibres by such conditions as pituitary tumours or craniopharyngeal tumours. Lesions of the optic chiasma cause optic atrophy visible with the ophthalmoscope.

(*d*) The Optic Tract

This contains the nerve fibres which correspond to the total opposite field of vision, and lesions in this structure cause defects of both the opposite fields of vision which are

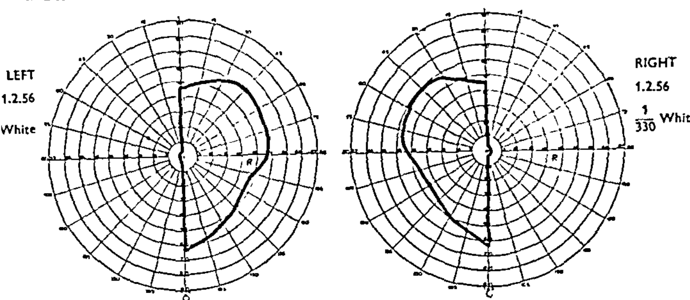


Fig. 76 Visual field charts of a patient showing left homonymous hemianopia

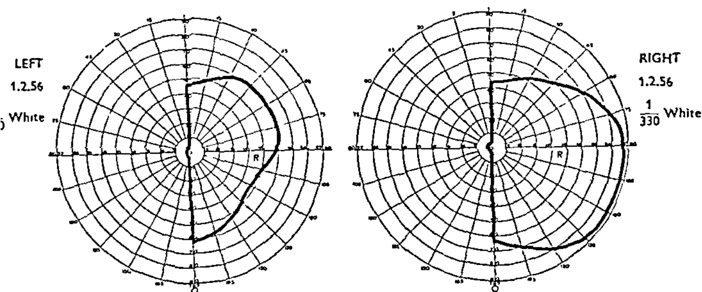


Fig 77. Visual field charts of a patient showing left bitemporal hemianopia.

called homonymous defects (Fig. 76). The fibres adopt an arrangement by which those of corresponding points of the two retinæ take up a state of apposition. This adjustment is not complete in the anterior part of the optic tract and lesions here cause a differing loss of the two fields, and are called incongruous defects in contrast to the more similar or more congruous defects which result from lesions in the posterior part of the optic tract and to the completely congruous defects caused by lesions of the optic radiations and occipital cortex. Lesions of the optic tract lead to optic atrophy.

(*e*) The Lateral Geniculate Body

The fibres from the ganglion cell layer of the retina pass in the optic nerve, optic chiasma, and optic tract and relay with the nerve cells of the lateral geniculate body, the fibres of which pass to the cerebral cortex. Localized lesions of this part of the brain, which are uncommon, cause homonymous congruous defects.

(*f*) The Optic Radiations

The fibres of the cells of the lateral geniculate body pass through the posterior part of the internal capsule to the optic radiations, in which the corresponding fibres of the same parts of both retinæ are in close association so that lesions cause similar or congruous defects. Lesions of the optic radiation do not cause optic atrophy.

(*g*) The Visual Cortex of the Occipital Lobe

The fibres of the optic radiation pass to the visual cortex of the occipital lobe. The visual cells are grouped around the calcarine fissure on the medial aspect of the occipital lobe. The upper lip of the calcarine fissure corresponds to the upper quadrant of the temporal retina of the same side and the upper quadrant of the nasal retina of the opposite side while the lower lip of the calcarine fissure corresponds to the lower quadrant of the temporal retina of the same side and the lower quadrant of the nasal retina of the opposite side. The posterior part of the calcarine fissure and the posterior pole of the brain correspond to the macular area. Lesions of the visual cortex do not cause optic atrophy.

(2) Higher Visual Pathways. Defects of the visual cortical communications may be characterized by various defects.

(*a*) Visual Agnosia

This is a loss of ability to recognize objects perceived, although the reception of sensory impressions is normal. Visual object agnosia is an inability to recognize common objects by sight although they can be seen, and they can be recognized by touch. This condition usually occurs in lesions close to, but not of, the area striata. Colour agnosia, which is the inability to recognise colours, although they are seen, and spatial agnosia which is the inability to appreciate the relative position of objects in space may also occur.

(*b*) Alexia (word blindness)

This is a loss of the ability to recognize words and it may be congenital or it may be acquired, and it may, in the latter case, be due to a lesion of the angular gyrus of the dominant hemisphere, or to a defect of the occipital region.

(*c*) Cerebral Distortion

Objects may be distorted (metamorphopsia), or enlarged (macropsia) or diminished (micropsia) as the result of cerebral disturbances.

Lesions which may Cause Defects of the Visual Pathway. A variety of causes may be responsible for disorders of the visual pathway.

(1) Injury (page 96)

(2) Inflammation

Inflammation of the meninges as well as of the brain itself may cause defects of the visual pathway.

(3) Neoplasms

Orbital neoplasms have been discussed (page 120). Neoplasms of the brain, both primary and metastatic may cause defects of the visual pathway. The visual defect may be the first symptom in some cranial tumours of primary type, but the primary growth has usually been recognized in the metastatic cases. Pressure from aneurysms occurring in the region of the circle of Willis must always be considered in the causation of lesions of the visual tract.

(4) Vascular Causes

Thrombosis of the intracranial vessels, and hæmorrhages from these vessels may cause deficiencies of the visual fields.

(5) Diseases of the Nervous Tissues

Tabes dorsalis may be associated with atrophy of the optic nerve fibres, and it seems that the process may sometimes affect the visual fibres in the region of the chiasma so that bitemporal lesions are seen. Disseminated sclerosis may affect any part of the visual tract from the optic nerve to the cortex, although most commonly it affects the optic nerve as the characteristic retrobulbar neuritis. Schilder's diffuse periaxial encephalitis is an uncommon condition which affects the occipital cortex and causes demyelinization of the visual nerve fibres and which may lead to complete blindness of cortical type.

Hypertension

Hypertension or the permanent rise of the blood pressure above what is considered to be normal may be primary, when it is called essential hypertension, or secondary, when it is due to renal disease, either nephritis, chronic pyelonephritis, polycystic kidney, renal calculus, or vascular lesions of the kidney, or to such conditions as coarctation of the aorta, polyarteritis nodosa, toxæmia of pregnancy or to that neoplasm usually arising in the adrenal body which is called a phæochromocytoma. The condition may remain benign both in essential hypertension and in all the types of secondary hypertension, or it may both in primary and secondary forms progress to the malignant form associated with extensive retinal changes and with renal deterioration. This usually proceeds to a fatal termination. Hypertension, it is generally recognized, is due to increased resistance of the peripheral circulation; there are many causes of this resistance. The level of the hypertension is satisfactorily assessed by measuring the level of the diastolic pressure.

Symptoms and Signs. The condition may be symptomless or there may be such symptoms as headache, visual defects, dyspnœa, and fatigue. The signs occur in the vascular system, and these become manifest particularly in the retina. These retinal changes have been classified by Keith, Wagener, and Barker into four grades which have important prognostic significations:

(*e*) The Lateral Geniculate Body

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low plasma proteins since the onset is often delayed until the volume of the blood has returned to normal, though its essential constituents and especially plasma proteins remain defective.

Symptoms and Signs. The visual defect, usually binocular, but sometimes uniocular, comes on in the majority of cases within 7 days of the commencement of the hæmorrhage. The retinal arterioles are found to be constricted, and this is associated with retinal œdema and with hæmorrhages and there is blurring of the optic disc. Optic atrophy develops and persists in association with considerable loss of the visual field. This field defect often affects the central area, and it has been suggested that the macular cells are more susceptible to the effects of hæmorrhage than are those of the rest of the retina. Considerable recovery of vision may occur in the succeeding months. It is found that the hæmorrhage has usually occurred from some disease process, such as a gastric ulcer or a diseased uterus, and that it is only rarely that it follows the hæmorrhage of trauma. History records that it has followed venesection. These cases are not common, but they are of such serious nature when they do occur that the condition should always be remembered in cases of hæmorrhage.

Treatment. The most necessary point is to stop the hæmorrhage and to give blood transfusions. There are solid grounds to support the belief that the prompt use of transfusions in all cases of hæmorrhage, and especially when the hæmoglobin falls to less than 50 per cent, will prevent this form of visual defect. It has been observed, however, that while transfusion seems to prevent this visual defect, it is not, after visual defect has occurred, an insurance against further loss, nor does it give any guarantee that recovery of vision will occur. It has been suggested that paracentesis of the anterior chamber of the eye should take place in these cases in order to restore, by lowering of the intraocular pressure, the circulation of the blood in the retina. There is, however, no evidence that this procedure has been outstandingly successful. Prevention by transfusion seems all important, and no treatment seems to be successful once loss of vision has occurred.

Leukæmia. This condition, which is characterized by an increase in the number of circulating white blood corpuscles, may cause small nodules, formed of the characteristic cells both *in the conjunctiva and in the orbit, and these may be the first manifestation of* the disease. Chloroma tumours occurring in the orbit of children have already been discussed (page 123). Leukæmia produces a characteristic retinopathy in which the fundus may present a grey appearance with dilated tortuous veins. Hæmorrhages may be present having characteristic white centres, and fluffy exudates may be seen. This retinopathy which is a sign of fairly advanced leukæmia may be accompanied by some infiltration of leukæmic cells into the choroid and even into the iris.

Hæmophilia. This familial disease leads to severe bleeding after injury, not only in the eyes and ocular adnexa, but in all parts of the body. Spontaneous hæmorrhages may occur in the orbit causing very marked proptosis, and bleeding may occur in the vitreous and under the conjunctiva.

Endocrine Exophthalmos

This condition occurs in association with thyrotoxicosis and it is one of the most outstanding signs of this condition. It is usually bilateral, but one eye may be more prominent than the other or the condition may occur in one eye before the other. This prominence of the eyes has given rise to certain classical signs, which include: (1)

Grade I. Slight narrowing or mild sclerosis of the retinal arterioles.

Grade II. More marked sclerosis of the retinal arterioles, with exaggerated light reflexes, irregularity of the lumen, and some compression of the arteriovenous crossings.

Grade III. Retinopathy with hæmorrhages, particularly of the flame-shaped type and exudates both of the fluffy cotton wool type and of the firmly circumscribed type. There is usually marked sclerosis or attenuation of the arterioles.

Grade IV. Similar condition to Grade III accompanied by measurable papillœdema.

It is assumed that Grade I includes those patients who have the best prognosis for survival, while Grade IV contains those with the worst prognosis for survival. Examination has shown that kidney damage is usually most advanced in those cases which show the fundus changes of Grade IV, but the immediate cause of the retinopathy is uncertain. There is some difference of opinion regarding the type of fundus changes which indicates the state of malignant hypertension. Some observers state that the presence of papillœdema is the characteristic factor, but the majority of workers consider that the presence of extensive retinal exudates and hæmorrhages indicates the malignant phase.

Treatment. The treatment of benign hypertension is symptomatic, and since such patients may remain symptomless for many years treatment may be unnecessary for a long period. The treatment of malignant hypertension may be undertaken by medical means by the use of hypotensive drugs such as hexamethonium bromide, or may be surgical by removal of the thoracic and lumbar sympathetic chains on both sides and by division of the splanchnic nerves. This surgical procedure causes an immediate diminution of the level of the blood pressure which, however, gradually rises again during the year which follows the operation. The surgical treatment causes improvement of headache and of other symptoms, but it is doubtful whether it prolongs life in any patient showing Grade III or Grade IV retinopathy at the time of operation. Both medical and surgical treatment cause an immediate clearing of the hypertensive retinopathy though vascular sclerosis, of course, remains unchanged.

Blood Diseases

Anæmia. This condition may be primary (pernicious anæmia, and iron deficiency anæmia) or secondary, when it is due to hæmorrhage from the stomach, duodenum, rectum, uterus, or other organs. Anæmia is always manifest in the eyes by pallor of the palpebral conjunctiva, demonstrable by depressing the lower eyelid. Hæmorrhages and exudates may be observed in the retina by ophthalmoscopic examination.

Hæmorrhage as a Cause of Optic Atrophy. It has long been recognized that blindness or severe visual defect may follow severe hæmorrhage. It seems that the visual loss is not related to the amount of blood lost, and that the vision is more likely to be affected in patients who have repeated hæmorrhages, than in those who suffer one big hæmorrhage. *Pathology.* The eyes of these patients do not become available for histological examination, but animal experiments have suggested that the ganglion cells and the nerve fibre layer of the retina are predominantly affected. This corresponds with the pathological findings in cases of obstruction of the central artery of the retina. The cause of the blindness is uncertain. Hæmorrhage into the sheaths of the optic nerve, retrobulbar neuritis, blood stasis causing retinal œdema and hæmorrhage, and thrombosis of the central artery of the retina have been postulated. It seems that the ischæmia of the retina must be the important cause, and it has been suggested that the damage is due to

CHAPTER III

EAR, NOSE AND THROAT

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THE *general surgeon* is concerned with *pain, swellings*, and *inflammation*, in and around the ear. It is of some relief to him to know that bone disease (mastoiditis) is very much less common than it was, thanks to early recognition of otitis media, and its rapid resolution with antibiotics. For *mastoiditis* is simply one of the complications of otitis media. It is caused by the spread of infection and inflammation from the middle ear cleft to the adjacent bone. To the aural surgeon, mastoiditis is osteitis, or osteomyelitis, in the bones of the base of the skull. The general surgeon will find that the signs and symptoms of mastoiditis are exactly the same as those for osteitis and osteomyelitis elsewhere. The same medical knowledge that has reduced the incidence and the serious complications of osteomyelitis elsewhere in the body, has had a similar effect on mastoiditis.

ACUTE OTITIS MEDIA

The middle ear cleft is a space in the thickness of the bone of the base of the skull. It has a communication with the nasopharynx, via the eustachian tube, and is closed off from the outside air by a fairly delicate membrane of skin, known to all of us as the ear drum, or tympanic membrane. The external auditory meatus, which enables us to see the drum, is just over one inch in length. Sometimes, a week or 10 days following the onset of an upper respiratory infection, the middle ear (which is lined by and developed from, the same moist, mucus-secreting, epithelium of the nasopharynx), becomes inflamed. Possibly the infection is the result of direct spread. It is usually due to a Gram-positive coccus, and is thought to belong to that group of cocci which follows in the wake of a virus infection of the upper respiratory tract. Otitis media often follows excessive blowing of the nose, when the fingers are gripping the nostrils. This habit of pressing on the nostrils when blowing the nose increases the positive pressure of the air in the nasal passages, and should be discouraged. It may cause the patient to force pus or sticky mucus past the valves of the eustachian tubes.

ACUTE OTITIS MEDIA COMPLICATING GENERAL SURGICAL CASES

The general surgeon in charge of patients immobilized in the supine position with a fractured femur, a spinal injury, or with any serious incapacitating disease, should realize that sticky, infected mucus tends to collect, in ill-health, in the nasopharynx. Where possible, movement, either passive or active, under a ward physiotherapist will minimize this risk. But the surgeon should always be on the look-out for otitis media if his immobilized patient contracts a head cold. Gargling, mouth washes, sitting up for short intervals, instructions in blowing the nose with the nostrils free, occasional inspection of the tympanic membranes with an auriscope, and systemic antibiotic treatment of

von Graefe's sign, which is characterized by a delay of the upper eyelid in following the movement of the eyeball from above downward; (2) *Stellwag's sign*, which is a decrease of the blinking reflex; (3) *Moebius' sign*, which is a weakness of convergence power. The prominence may be so great that the cornea cannot be completely covered by the eyelids, and the cornea may ulcerate and even perforate. The cause of the exophthalmos is uncertain but it appears to be associated with some increase of the orbital fat and with some atonicity of the extraocular muscles. Pressure upon the eyeball, in these cases, can force it back into the orbit, for so long as the pressure is applied.

The exophthalmos of thyrotoxicosis differs from another type of exophthalmos which may occur after partial thyroidectomy for toxic symptoms, and is thought to be due to a hypersecretion of the thyrotropic hormone of the pituitary gland. It appears that, due to defective thyroid secretion, this hormone which stimulates the activity of the thyroid gland is produced in excessive amount. This causes a lymphocytic reaction and organization in the tissues of the orbit, and the eyeball is protruded. This protrusion of the eyeball cannot be overcome by backward pressure on the eyeball, because the infiltration of the orbital tissues is very dense. This may be associated with chemosis and with œdema of the eyelids, and often there is some defect of ocular movement. The condition may progress until the cornea becomes ulcerated from exposure.

Treatment. Thyrotoxic exophthalmos is treated by thiouracil or by partial thyroidectomy. The proptosis usually improves with treatment, but it may not disappear entirely, even if all toxic symptoms have ceased. Thyrotropic exophthalmos has been treated with thyroid, thiouracil, and œstrogens, and by radiotherapy both to the pituitary gland and to the orbit. The more severe cases require decompression of the orbit which is usually carried out by a transcranial approach and by removal of the roof and outer side of the orbit. Tarsorrhaphy should be done at this time. These cases appear to be self-limiting and provided corneal scarring can be avoided the ultimate prognosis for vision is satisfactory. The residual proptosis may be minimal.

The author wishes to thank the Medical Illustration Department of the Institute of Ophthalmology for many of the illustrations reproduced in this chapter.

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the otitis media at the earliest possible moment, will save much discomfort, suffering, and damage to hearing. Tiny infants immobilized and dehydrated from intestinal infections are notoriously prone to otitis media. Children immobilized for long periods in plaster are also liable to ear infections in this way, particularly if there is a strong family history of otitis media. *Acute otitis media* is easily recognized by deep seated earache and pyrexia, redness of the tympanic membrane, associated with, as a rule, some intercurrent nasopharyngeal infection.

TRAUMA TO THE EAR COMPLICATING GENERAL SURGICAL CASES

Surgeons should accept *trauma* to the ear as a reasonably likely complication of *fractures of the base of the skull*, and of wounds associated with *blast*. During the immediate routine examination of a patient suspected of these injuries, the tympanic membranes should *always* be examined with an auriscope, whether or not blood or C.S.F. is escaping from the external auditory meatus. An auriscope helps because it magnifies. Trauma to the tympanic membrane is characterized by hæmorrhages and tears in the membrane. The surgeon should apply his knowledge of the physical signs of trauma elsewhere in the body to the small, but easily magnified, tympanic membrane.

INFECTION SUPERVENING UPON TRAUMA

It is of very great importance to try to prevent infection reaching the damaged ear because otherwise complications such as deafness, meningitis, or mastoiditis may result. The meatus of the ear should be protected with a clean cotton wool filter, and adequate antibiotic treatment given (if this has not already been ordered for the patient's other injuries). The damaged tympanic membrane, and the external auditory meatus should be left alone, and interfered with as little as possible. Most cases of trauma heal well, and the hearing tends to improve as resolution and healing take place. The tympanic membrane should be examined from time to time to observe progress, and notes made of the signs when first seen, and the subsequent changes, not only for the sake of the patient's immediate health, but in order to record facts that may be useful to medical colleagues should the questions of pension or insurance claims result. The help and advice of an aural colleague should be sought whenever possible

CHRONIC OTITIS MEDIA COMPLICATING GENERAL SURGICAL CASES

Chronic otitis media will probably have been present long before the general surgical disease for which the surgeon has admitted the patient It is usually discovered on routine examination, because the patient tells the house surgeon that he has an ear discharge. Chronic purulent otitis media may be important to the orthopædic, thoracic, or abdominal surgeon, or to the neurosurgeon and obstetric surgeon, because it indicates the presence of intercurrent infection. Often slow bone caries may be present in the middle ear, and Gram negative organisms of intestinal origin, like B. coli, B proteus, or B. pyocyaneus, or the Gram positive and rather formidable staphylococcus pyogenes, may be active. Such cases should be assessed for the general surgeon by his colleague, the aural surgeon. Chronic otitis media may, or may not, have a serious bearing on the patient's general surgical condition, or on the proposed operation elsewhere on the body.

It all depends upon whether there is *progressive disease* in the ear, or whether the purulent discharge is simply due to *dermatitis*. Sometimes there is an old perforation of the tympanic membrane, with only a moist mucus discharge, and no evidence of infection.

SIGNS OF CHRONIC OTITIS MEDIA

The signs of chronic otitis media are usually limited to a purulent discharge, without any pain. There may be some hearing loss, but whilst the patient is young, and the discharge is present, he often hears surprisingly well. Later in life, *deafness* may be a real handicap both to the patient, and to the general surgeon treating him for some other complaint.

DEAFNESS IN PATIENTS UNDER TREATMENT FOR SOME GENERAL SURGICAL DISEASE OR INJURY

Patients' deafness may be a great handicap to the general surgeon, and to the nursing staff. *Hearing aids* should be readily available nowadays in all big hospitals, or can be obtained from Hearing Aid Distribution Centres. The general surgeon must realize what an enormous boon hearing at normal conversational level will be to his deaf patient, and how much it will help his nurses in their onerous and difficult tasks. This is especially important in bed adjustment for incontinence, questions about the comfort or pain of plaster casts, splints, and wounds; normal micturition and defæcation when the deaf person is confined to bed; treatment to prevent bedsores; explanations, encouragements, words of hope; exercises and rehabilitation. To enable a severely deaf patient, facing a long period of confinement to his bed, to hear television or wireless, and the simple exchanges of visiting relatives or friends at normal conversational level, must be an enormous help towards recovery. Deafness condemns us to loneliness, and sad misunderstandings, and if a hearing aid can help, it should be ordered. The general surgeon should ask his aural colleague if a hearing aid would help. Sometimes the deafness may be due to severe cochlear destruction, and then, unfortunately, a hearing aid may be useless. The point to remember, however, is that to most deaf patients a hearing aid will give useful hearing, and a few all-purpose sets, kept up-to-date and in working order, should be available in all hospitals for deaf general surgical (and medical) patients. It should be the sister's responsibility to see that those hearing aids are returned to store when the patient leaves the ward. The aural surgeon can later arrange for proper estimation of the hearing loss, and the issue of a personal moulded ear piece, and suitable aid. Sometimes the aural surgeon may be able to restore really useful hearing by operation, and so enable the very deaf patient to do without a hearing aid.

INTRACRANIAL COMPLICATIONS OF OTITIS MEDIA

Complications depend upon the anatomical structures adjacent to the source of infection. In otitis media the infection is in the bone of the base of the skull (the middle ear cleft and tympanic antrum).

In the early stages, an infection of the middle ear is limited to the mucous membrane with which the middle ear is lined. If this is destroyed by disease, the infection will spread to bone. First an osteitis, then possibly an osteomyelitis may follow if the infection is untreated. The spread of infection in bone depends upon the type of organism present

the otitis media at the earliest possible moment, will save much discomfort, suffering, and damage to hearing. Tiny infants immobilized and dehydrated from intestinal infections are notoriously prone to otitis media. Children immobilized for long periods in plaster are also liable to ear infections in this way, particularly if there is a strong family history of otitis media. *Acute otitis media* is easily recognized by deep seated earache and pyrexia, redness of the tympanic membrane, associated with, as a rule, some intercurrent nasopharyngeal infection.

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CHRONIC OTITIS MEDIA COMPLICATING GENERAL SURGICAL CASES

Chronic otitis media will probably have been present long before the general surgical disease for which the surgeon has admitted the patient. It is usually discovered on routine examination, because the patient tells the house surgeon that he has an ear discharge. Chronic purulent otitis media may be important to the orthopædic, thoracic, or abdominal surgeon, or to the neurosurgeon and obstetric surgeon, because it indicates the presence of intercurrent infection. Often slow bone caries may be present in the middle ear, and Gram negative organisms of intestinal origin, like B. coli, B. proteus, or B. pyocyaneus, or the Gram positive and rather formidable staphylococcus pyogenes, may be active. Such cases should be assessed for the general surgeon by his colleague, the aural surgeon. Chronic otitis media may, or may not, have a serious bearing on the patient's general surgical condition, or on the proposed operation elsewhere on the body.

The surgeon should remember his anatomy. He will be working downwards through a small triangle in the bone behind the ear, bounded in front by the bony meatal wall, above by the zygomatic ridge, and posteriorly by an imaginary line running in the direction of the mastoid process.

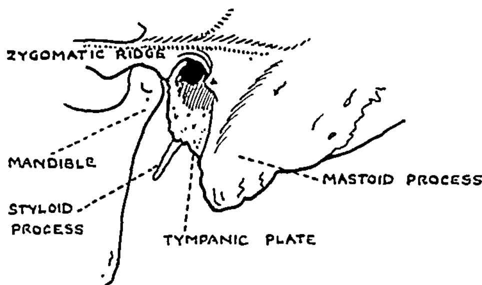


FIG 78 Anatomy of bone for operations on the ear.

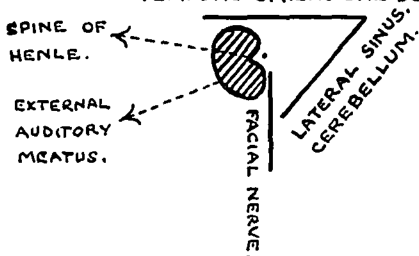


FIG 79 Diagram showing important anatomical relations of approach to tympanic antrum

Above will lie the dura of the middle fossa; posteriorly, the lateral venous sinus and the cerebellum; anteriorly the facial nerve. The line for the facial nerve is roughly the fissure between the mastoid process and the tympanic plate. It lies on the medial wall of the aditus, in a shallow bony canal, where it is easily injured, between the horizontal semicircular canal and the oval window. The medial wall of the tympanic antrum is a good guide to its depth at this point. In a simple drainage operation of the tympanic

and the rapidity with which the infection spreads through the bone. Meningitis of otitic origin is almost always streptococcal or pneumococcal.

(*a*) Rapid Spread of Infection

(1) Acute mastoiditis, rapid spread through cellular bone with osteomyelitis.

(2) Spreading meningitis, if infection reaches meninges of tempero sphenoidal lobe above, or sub tentorial cerebellum posteriorly.

(3) Septicæmia, if infection spreads rapidly to lateral venous sinus or along petrosal venous sinuses.

(4) Facial paralysis, acute labyrinthitis, deafness.

(*b*) Slow Spread of Infection

(1) Chronic bone disease, sclerosis, often associated with cholesteatoma.

(2) Bone abscess; extra dural abscess; brain abscess—temporo-sphenoidal or cerebellar.

(3) Thrombophlebitis of lateral venous sinus or petrosal sinuses; with spread via jugular vein, heart, and pulmonary artery to lungs, broncho-pneumonia, lung abscess.

(4) Facial paralysis due to erosion of bony canal. Erosion and destruction of labyrinth and cochlea.

OPERATION FOR ACUTE MASTOIDITIS (Figs. 78, 79)

Fortunately this is a comparatively rare operation today, although before the days of penicillin (first available for civilians in Great Britain 1945) an operation to drain the pus from the tympanic antrum and mastoid air cells was the only treatment for acute mastoiditis.

If the general surgeon is called upon to perform such an operation he should refresh himself with regard to the anatomy of the ear. The operation is one of simple drainage of an abscess in bone in a singularly dangerous place. The aim of the surgeon is to open the tympanic antrum. That alone will often suffice, although if he is experienced, the surrounding infected mastoid air cells can also be opened.

The surgeon makes an incision behind the pinna. If he is inexperienced the incision may be made right down to bone following a line just posterior to the post aural sulcus. It is usual for the aural surgeon to make his incision in the post aural sulcus to avoid subsequent scarring. The periosteum is incised and raised from the mastoid bone, until the rounded circumference of the bony external auditory meatal wall is identified. Above will lie the bony ridge which is the posterior continuation of the upper border of the zygoma. This ridge runs backwards from the upper border of the bony meatus. In the posterior bony wall of the meatus is a well defined spine with a dimple in its centre. This is called the Spine of Henle. Directly posterior to this is the tympanic antrum. Today the aural surgeon uses a rotating electric cutting drill for his bone work, and works with magnification X 2, using saline and suction to remove bone dust. The tympanic antrum is exposed, pus sucked out, and any infected bone cells opened and similarly treated. Systemic penicillin is ordered, and, if advisable, the wound may be drained. It is common practice today to regard the operation as one of simple drainage of an abscess in bone, and it is usual to close the wound, and to rely on penicillin to assist full resolution. If much cellulitis is present, however, a drain may be inserted.

(3) Pain in Front of the Ear

Arthritis mandibular joint.
Parotitis.
Mixed salivary tumour.

(4) Pain in the External Meatus

Furuncle or fissuring from dermatitis.
Foreign body.
Impacted wax.

(5) Pain in the Pinna Itself

Hæmatoma, trauma.
Dermatitis, cellulitis, eczema.

(6) Pain in the Middle Ear

Otitis media.
Blast injuries to the tympanic membrane.
Cerebral complications of otitis media.

REFERRED PAIN TO THE EAR

Common Examples

(1) **Dental.** Via inferior dental nerve to auriculo temporal (particularly from wisdom teeth).

(2) **Pharyngeal.** Tonsillitis, quinsy, carcinoma; via glosso-pharyngeal nerve to tympanic plexus.

(3) **Tongue.** Carcinoma, and painful ulcers. Pain referred via lingual nerve to auriculo temporal nerve.

(4) **Laryngeal.** Laryngitis, carcinoma; via superior laryngeal of vagus to auricular branch of vagus.

THE EAR IN FACIAL PARALYSIS

(1) Bell's Palsy

There is often initial pain in the ear and face. Cases have been observed of transient redness of the tympanic membrane on the first day of paralysis. Most cases of Bell's Palsy recover rapidly, but in those cases where the nerve has had to be decompressed, inflammation and œdema of the nerve is commonly observed at operation under magnification. The facial paralysis is typical of a lower motor neuron lesion, and affects both upper and lower parts of the face.

(2) Fractured Base

The facial paralysis is sometimes accompanied by the escape of blood, or C.S.F. from the meatus, or tears may be seen in the tympanic membrane. If the facial paralysis, which is of a lower motor neuron type, comes on simultaneously with the head injury, the surgeon should suspect that the nerve may have been nipped between fissures of

antrum, it is unwise to probe through the aditus into the middle ear, because it is very easy to dislocate the short process of the incus when doing this.

The aural surgeon undertakes many operative procedures on the ear today, for the removal of tumours, the cure of chronic infection in the middle ear, relief of deafness, decompression of the facial nerve, fenestration of the labyrinth. He uses high magnification and an electric cutting drill, and his knowledge of the exact anatomy of the ear is an essential requirement. The general surgeon would be unwise to undertake any of these procedures unless he feels adequately and justifiably competent. He would be wise to confine himself otherwise to the simple drainage of the tympanic antrum described above.

PAIN IN AND AROUND THE EAR (Fig. 80)

(1) Pain Behind the Ear

Mastoiditis, periostitis. Pain on deep pressure; swelling, œdema, sometimes redness of skin; evidence of inflammation of tympanic membrane or discharge from middle ear.

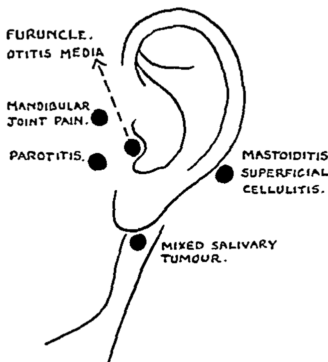


FIG. 80. Pain in and around the ear.

Superficial inflammation in lymph node, or cellulitis from furuncle of external auditory meatus. Superficial hyperæsthesia as compared with pain on deep pressure in bone disease. Tympanic membrane will be intact. Furuncle will possibly be seen in meatus. No otitis media. Pinna painful when moved. (In otitis media the infection is in the bone of the middle ear cleft and movement of the pinna gives no pain.)

(2) Pain Below the Ear

Mumps, often early, before any swelling.
Cervical adenitis.
Mixed salivary tumour (mixed parotid tumour).

(3) Pain in Front of the Ear

Arthritis mandibular joint.
Parotitis.
Mixed salivary tumour.

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Furuncle or fissuring from dermatitis.
Foreign body.
Impacted wax.

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displaced bone, or torn. He would do well to think of decompression if recovery is not speedy, and if the general condition of his patient allows it.

If the paralysis comes on a few hours or days subsequent to the time of the skull injury, the paralysis may be due to œdema or hæmorrhage, causing pressure on the nerve, and recovery should be expected, and hoped for.

The localization of the injury can be established with accuracy if the patient can co-operate. Involvement of other cranial nerves will serve as an additional guide.

(1) Chorda Tympani

If sensation of crude taste to sugar and salt in the anterior two-thirds of the tongue is absent, the lesion must be above the point where the chorda tympani nerve leaves the facial nerve (about 6 mm. above the upper apex of the stylo mastoid foramen). If the facial nerve is paralysed and the chorda tympani taste sensation is intact, the lesion is probably outside the base of the skull, or at any rate below the point where the chorda tympani leaves the facial nerve.

(2) Lacrimation

If the eye is dry on the paralysed side, the lesion may be above the geniculate gangleon (great superficial petrosal nerve). If lacrimation is present, the lesion is probably distal to the geniculate gangleon. Fine palatal taste sensation will either be absent or present, according to whether the same nerve is injured or not.

(3) Hyperacusis

If the lesion is above the point where the nerve to stapedius comes off the facial, at about the level of the oval window, disturbance to hearing may take place. This sign is not very conclusive, but the two preceding signs are very helpful indeed to the surgeon, and save much needless exploration and damage.

NOSE BLEEDING

Nose bleeding is fairly common in the elderly; can occur at any period of adult life; is fairly common in children about puberty, but is practically never met with in infancy.

(a) LOCAL CAUSES

(1) The commonest cause of *bleeding from the nose* is probably *inflammation of the nasal epithelium* with exposure of the underlying blood supply by shallow ulceration. To the general surgeon, hæmorrhage from the stomach, or intestine, is understandable, and indicates a similar process. Bleeding from the nose is immediately visible. The blood leaks from the nostril, or is spat out from the nasopharynx. Bleeding from the stomach and intestine is hidden unless severe. Consequently nose bleeding is more common because even very small losses of blood are recorded, whereas these pass unnoticed in the bowel.

Nose bleeding commonly follows *Upper Respiratory Infections*, and sinusitis. People suffering from chronic rhinitis or naso-pharyngitis are rather prone to find streaks of blood on their handkerchiefs.

(2) *Trauma* is a common cause of nose bleeding.

Displacement of the nasal bones or septum can tear the vascular mucosa inside the nose. Trauma due to surgical therapy must be included.

(3) *At High Altitudes* epistaxis is common.

(4) *Foreign Bodies* cause ulceration of the mucosa which tends to bleed.

(5) *Malignant Growth* in the nasal passages, ethmoid air cells, or maxillary sinuses may have, as its very first suspicious sign, a unilateral, blood stained nasal discharge.

(6) *Dilated blood vessels* on the septal mucosa just inside the vestibule of the nose, occur commonly in young and old people. These dilated vessels are due to obstruction at the incisive foramina in the floor of the nose, where terminal branches of four arteries, and four veins, meet (sphenopalatine, great palatine, superior labial, anterior ethmoid). They cause irritation, and may be picked with the finger, or bleed from slight inflammation. They become dilated, and visible, because past forgotten trauma has displaced the lower border of the quadrilateral cartilage of the septum from its insertion into the gutter of the vomer, thus obstructing the incisive foramina (Fig. 81).

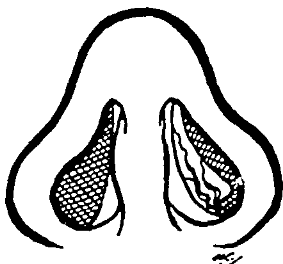


FIG. 81 Dilated blood vessels on nasal septum

(*b*) GENERAL CONDITIONS CAUSING EPISTAXIS

(1) **Hereditary multiple Telangiectasia.** The characteristics are aggregations of dilated small vessels, initially minute, but gradually increasing in size. They are scattered over the mucosa of the nose, mouth, and pharynx, and over the skin of the face and arms. These defects are not present at birth, but develop later, often in middle life. Cases vary in severity from complete harmlessness, to a cause of early death.

(2) **Hypertension, Apoplexy, Arterio-sclerosis.** Old people bleed more continuously from the nose than do the young. If blood-vessels are sclerotic they lose their powers of retraction and contraction, which are essential factors in hæmostasis. Physiological clotting (prothrombin + ionic calcium + thromboplastins from ruptured platelets or damaged tissue = thrombin; thrombin + fibrinogen from the liver = fibrin, the clot) can only take place if there is stasis. In the young person, contraction of the capillaries and arterioles follows shortly after injury, and it is this vital contraction which produces temporary hæmostasis and allows time for the formation of a strong fibrin clot. After injury, the damaged capillaries remain shut for from 20 minutes to 2 hours, and they resist distension from internal pressures up to 100 mm. of mercury. In old people the blood-vessels sometimes cannot contract, and so the application of temporary continuous firm pressure from without is needed.

(3) **Any of the Anæmias.** Leukæmias, Aplastic Anæmia, Pernicious Anæmia.

(4) **Blood Diseases.** Hæmophilia, Thrombocytopœnic Purpura.

(5) **Hodgkin's Disease.**

(6) **Hepatic Cirrhosis.**

(7) **Scurvy, Vitamin K deficiencies.**

(8) **Rheumatic Mitral Disease.**

(9) **Diphtheria, Typhoid Fever.**

(10) **Vicarious Menstruation.** Nose bleeding in young people at the time of the menstrual flow.

(11) **Upper Respiratory Infections.**

TABLE

BLOOD CHANGES AND THEIR CAUSES IN RELATION TO EPISTAXIS

Cause	Blood Changes
Anæmia due to chronic blood loss .	The changes in the blood are the result of the epistaxis, not the cause of it. Decrease in hæmoglobin. Decrease in red cells. Low colour index.
Aplastic anæmia	Blood platelets decreased nearly always. Decrease in red cells and leucocytes.
Thrombocytopenic purpura .	Coagulation time usually normal (up to 6 minutes, Dale and Laidlaw's method) Bleeding time prolonged (normal 2–4 minutes). Platelets reduced. Spontaneous skin hæmorrhages
Hæmophilia .	Rare cause of epistaxis Patient comes with a history of bleeding into joints Hæmorrhages follow injury Coagulation time prolonged—may be from 5 to 100 times the normal. Platelets normal (250,000–500,000 per cubic millimetre).
Leukæmias . .	Great increase leucocytes. may be 500,000 per cubic millimetre Platelets often decreased. Enlarged spleen In acute leukæmia, hæmorrhages into the skin.
Hodgkin's Disease	Hypochromic anæmia, lymphadenopathy.

ARREST OF NOSE BLEEDING

(1) This is sometimes (often fortunately) *spontaneous*. The power of the circulating blood to remain fluid in the veins and arteries, and to clot conveniently on rupture of a blood vessel, is one of the most amazing phenomena of human physiology. Without this property the surgeon would be helpless and hopeless, for, though readily accessible vessels may be ligated, capillaries must clot.

(2) Simple *pressing of the nostrils together* will stop bleeding from the anterior third of the nasal passages. It is sometimes possible to see the bleeding area.

(3) Bleeding from the *deeper nasal passages* can be very troublesome because it may be impossible to locate the bleeding point The inferior turbinate is very vascular; beneath its columnar epithelium are large arterio-venous blood spaces. There are similar blood spaces in the middle turbinate although these are not so numerous.

The arterial supply to the nose is mainly from the *sphenopalatine artery* which enters the nose through the spheno-palatine foramen, and supplies both septal and lateral aspects of the posterior two-thirds of the nasal passages. The sphenopalatine artery is the continuation, in the nose, of the maxillary artery. The anterior one-third of the nasal passage is supplied by the *anterior and posterior ethmoidal arteries*, which enter the roof of the nasal passages through the cribriform plate of the ethmoid along with the ethmoidal nerves. The anterior and posterior ethmoidal arteries are branches of the ophthalmic artery.

Bleeding from the nose may come from the nostrils, or may run down into the back of the throat where it can be spat out or swallowed. Provided the nasal passages are not obstructed, generally speaking, bleeding areas lying anteriorly will tend to lose blood from the *nostrils*, whereas bleeding areas in the posterior parts of the nasal passages will tend to lose blood into the *nasopharynx*. Post nasal bleeding can be very important in an unconscious patient.

Vomited Blood may be dark if there has been time for interaction with the hydrochloric acid of the stomach, but in severe hæmorrhage, the secretion of stomach hydrochloric acid is inhibited, and vomited blood may remain bright long after it has been swallowed.

Hæmoglobin Estimation

During the early stages of hæmorrhage, the blood volume will be reduced, and conserved by contraction of peripheral capillaries and arterioles. The hæmoglobin percentage will be normal because the blood density is the same. Later fluid is attracted into the blood vessels from the tissues, and the hæmoglobin percentage will correspondingly fall. A normal hæmoglobin percentage in the early stages of hæmorrhage must not entice the surgeon into a state of false security.

Simple Packing of the Nasal Passages with continuous gauze will arrest most nasal bleeding. We prefer to pack first with gauze wrung out in 5 per cent cocaine hydrochloride solution, and then to remove the gauze in 2 or 3 minutes and replace it with oily gauze worked up with some suitable antibiotic The cocaine is both analgesic, and a most reliable vasoconstrictor. Adrenaline is a good vasoconstrictor but produces a vasodilator action about an hour after its application, and may restart hæmorrhage. Adrenaline has no analgesic effect. Provided the patient does not swallow drops of the cocaine solution there will be no unpleasant reaction to this drug. The oily gauze prevents disturbance of fibrin on removal later. The antibiotic inhibits organisms from culturing in the obstructed nasal passages. Provided there is no generalized blood disease, simple stasis by gentle pressure of gauze in the nasal passages will suffice to cause fibrin to form, and so arrest hæmorrhage.

Blood Transfusion

The general condition of the patient will guide the surgeon. The volume of blood lost should be estimated. Here it is important for the nursing staff to keep a record of this as far as is humanly possible Where the blood loss has been fairly severe, the patient's blood should be typed and cross matched as a precautionary measure. Blood can then be ordered as necessary. Blood transfusion replaces lost constituents of the blood, and of itself encourages hæmostasis. Loss of blood makes the patient pale and restless. Transfused blood brings peace. It never increases the bleeding. The volume of blood to be

given depends on the volume of blood lost and the condition of the patient. It need never equal exactly the total volume lost.

Control of Severe Nose Bleeding

If the bleeding is not easily seen and controlled, and particularly if it is coming from the posterior, or sphenopalatine, area of the nasal passages, we at the London Hospital use *splints* which have proved very effective. A silk covered splint is inserted through each nostril, flat against the septum. The splint is long enough to fill the airway from the nostril in front to the posterior nares behind, and it must protrude into the nasopharynx for about half an inch. Local analgesia is used to enable this to be done; this is applied by gentle packing with cocaine gauze, before introduction of the splints, as described above. These splints are made of eight thicknesses of gauze, folded to form a flat roll, 1 *in. wide, one eighth of an inch thick,* and about 6 in. *long.* Oiled silk is folded over the gauze. A stout black waxed thread is sewn into one end (Fig. 82). The surface marking of

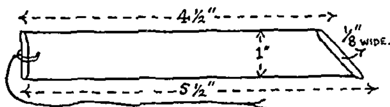


Fig. 82 Nasal splint.

the posterior nares is estimated by feeling for the tip of the coronoid process of the mandible. The splint is cut to the required size, so that the end of the splint will pass through the posterior nares. This is important because in this way it effectively blocks the posterior nares and prevents bleeding into the nasopharynx. The supreme simplicity of this method has much to recommend it. The surgeon is always concerned with the possibility of continuing post nasal bleeding, and may not have any of the more ingenious rubber bag instruments handy. Rubber bags can, unfortunately, deflate, or perish in store, but these simple splints can always be made up on the spot by any nurse, with *materials available in any hospital at any time.* Post nasal packs are uncomfortable, and may cause distress. The patient is restless enough from blood loss. Furthermore post nasal splints have to be taken out from behind, and this again can be a difficult thing to do. The use of the splints described *solve many problems and* can be *inserted* and removed through the nostrils from the front. They are secure because the patient cannot swallow them. They are comfortable. They can be smeared with antibiotic paste and so prevent infection and secondary hæmorrhage.

After the splints are in place, *ribbon gauze,* worked up in an oily solution, with appropriate antibiotics, is packed into the nasal passages lateral to the splints. It is convenient to have a nurse hold the splint steady against the columella with a pair of fine artery forceps, to keep it firm whilst the lateral packing is done. *A suitable mixture* for oiling this gauze is, liquid paraffin two millilitres, sulphathiazole one gramme, penicillin fifty thousand units; but any simple antibiotic paste preferred can be used.

The splints are tied across the columella. The ends of the ribbon gauze, lateral to the splints, are also tied across the columella.

Morphine is given for rest and sleep, and to promote stasis, once the bleeding has been successfully stopped and the blood volume restored. It is most unwise to give morphine if a second anæsthetic is contemplated, after failure to arrest hæmorrhage at the initial operation.

The splints and lateral gauze are inserted in the following order:

(1) Local analgesic packing.
(2) Withdrawal in 2 or 3 minutes.
(3) Insertion of splints.
(4) Insertion of lateral gauze.

They are removed in reverse order:

(1) Removal of lateral gauze in 24 hours. If no bleeding, then
(2) Removal of splints.

If bleeding recurs after removal of the lateral gauze, the nose should be repacked with fresh lateral gauze, the nasal splints remaining in place.

Secondary Hæmorrhage from the Nose

Secondary hæmorrhage usually takes place about 6–10 days after an injury, or operation, and is due to infection. There is usually some slight pyrexia and anorexia. Secondary hæmorrhage is thought to be due to systemic, and not local, causes. It is known that the streptococcus produces a fibrin destroying enzyme—streptokinase—and this, circulating in the blood, dissolves the fibrin clot. Systemic antibiotics prevent and cure most nose bleeding from this source, but if the bleeding is severe, packing to prevent further blood loss is indicated, but penicillin or some other suitable antibiotic must be given systemically to allow firm fibrin to form.

Nose Bleeding during Anæsthesia

The passage of an endotracheal catheter through the nose may cause considerable bleeding if there is pre-existing inflammation of the nasal mucosa. Instillation of vasoconstrictor/analgesic agents will greatly reduce this risk where inflammation is known to be present, to the great advantage of anæsthetist and surgeon. These should be instilled whilst the patient is conscious, and great care taken to prevent the patient from swallowing the drugs. The following is a useful vasoconstrictor/analgesic mixture, but it must be mixed immediately before use or precipitation will take place.

Adrenaline 1 in 1000 1·2 ml.; cocaine hydro chloride 10 per cent 2·4 ml.; sod. bic. 1 per cent 2·4 ml.

Severe epistaxis can be quite disconcerting. A little blood can make an awful mess of bedclothes and towels A lot of blood looks terrifying. The patient is anxious because he thinks he is going to bleed to death. In hospital it is bad enough. At home it is worse. The relatives are filled with apprehension, and often worn out from unavailing efforts to stop it. The surgeon is often anxious too, because he is not sure that he can stop it either. If the surgeon uses the simple technique described above, he will have confidence in his ability to control any epistaxis, however severe. There is an old wives' tale that bleeding from the nose will always eventually stop, as the blood pressure falls with excessive blood loss. This is not true. Persistent bleeding over a number of days can be just as dangerous as one severe dramatic hæmorrhage.

Example. A fine young man had been bleeding from the nose for over a week before the author of this section was sent for. Although the patient was visited promptly, he was dead before help could arrive. He had been swallowing blood, and in the turmoil of modern war conditions, his danger had perhaps not been fully appreciated. At autopsy, no other cause for his death was found.

THE LARYNX

Acute laryngitis is fairly common. The patient speaks with a forced whisper, and says that she has lost her voice. There is usually a history of a recent upper respiratory infection. Acute laryngitis simply means an inflammation of the larynx. Symptoms depend, of course, upon the severity of the inflammation. This is usually mild, and apart from the loss of voice, the patient is none the worse.

On the other hand, the laryngitis may be accompanied by tracheitis and bronchitis. Speaking may cause actual pain. Even the passage of air through the larynx may cause discomfort, particularly cold air. Coughing may be painful, because the cords have to be adducted when expelling mucus from the respiratory passages.

Occupational Causes. People who tend to pitch their voices, in an effort to throw the sound to an audience, are rather more prone to laryngitis—teachers, lecturers, preachers, actresses. Excessive smoking of cigarettes and exposure to occupational air pollution tend to cause irritation to the pharyngeal, laryngeal, and bronchial mucosa, and these patients are rather prone to laryngitis.

Shouting for long periods at football matches, race courses, or on great emotional occasions, can strain the vocal cords, causing temporary loss of voice. This is really a type of traumatic laryngitis, and the inability to speak is probably a protective reflex.

Hysterical Aphonia. People with emotional personalities tend to lose their voices easily. There is usually some slight initial physical cause for the aphonia—like a mild forgotten upper respiratory infection—but when the vocal cords and larynx are examined, no cause for the loss of voice can be found. There is no paralysis and no inflammation to be seen. The voice is weak and the patient speaks in a whisper. There is often a history of previous attacks of aphonia. The voice always returns to normal between attacks.

Œdema of the Larynx

This is nearly always due to a combination of two factors—inflammation and trauma. If there is pre-existing acute inflammation of the larynx, even such slight trauma as the passage of an endotracheal tube can cause acute œdema. Respiratory embarrassment is nearly always at the cricoid ring because this is the only part of the respiratory tract where the cartilaginous ring is complete and allows of no expansion. The timely use of antibiotics will obviate the necessity for tracheotomy.

Chronic œdema of the larynx is seen round tuberculous ulcers, or where carcinoma has obstructed the proper exchange of tissue fluid. Chronic œdema seems to affect mainly the arytenoid eminences, and considerable thickening sometimes follows radiotherapy.

Clinical Precaution

Any doctor, general surgeon, or general practitioner should become suspicious of *any hoarseness that has been present for 3 weeks.* In that time, any acute laryngitis should

be recovering. The risk of early carcinoma of the vocal cords is always present in the middle-aged—especially in men. In its early stages, carcinoma in this position is very definitely curable. The larynx of any patient with a persisting hoarseness after 3 weeks, should be seen by a competent examiner.

The general surgeon should remember that inflammations tend to recover, but any tumour, however benign, on a vocal cord, will persist, and continue to produce changes in the voice.

CAUSES OF HOARSENESS IN ADULTS

(1) Laryngitis, Acute

- (*a*) Non specific.
- (*b*) Streptococcal.
- (*c*) Diphtheritic.
- (*d*) Poisonous gases.
- (*e*) Very intense heat.

(2) Laryngitis, Chronic

- (*a*) Excessive smoking.
- (*b*) Occupational air pollution.
- (*c*) Tuberculosis.
- (*d*) Syphilis.

(3) Neurosis

- (*a*) Hysteria.
- (*b*) Emotional anxiety.

(4) Stenosis

- (*a*) Traumatic.
- (*b*) Fibrous secondary to
  - (i) inflammation.
  - (ii) caustics.
  - (iii) intense heat.
- (*c*) Congenital webs.

(5) Trauma, Internal

- (*a*) Shouting.
- (*b*) Singing with strain.
- (*c*) Post-operative.

(6) Trauma, External

- (*a*) Boxing.
- (*b*) Judo.
- (*c*) Suicides.
- (*d*) War wounds.

(7) Paralysis: Unilateral, Bilateral

(*a*) Disease or injuries to the nucleus ambiguous.

(i) Medullary poliomyelitis (common).
(ii) Brain injuries to the medulla.
(iii) Tumours of medulla.
(iv) Tabes, multiple sclerosis, syringomyelia.

(*b*) Polyneuritis; toxic, virus.
(*c*) Local myopathies; myasthenia gravis.
(*d*) Pseudo-paralysis, due to fixation by local spreading malignant growths.
(*e*) Recurrent laryngeal nerve paralysis; lesions in

(i) the lower neck.
(ii) apices of either lung.
(iii) near the aortic arch.

(8) Tumours, Non-malignant

(*a*) Polypi.
(*b*) Cysts.
(*c*) Papillomas.
(*d*) Chondromas of cricoid cartilage.
(*e*) Fibromas, neuromas, angiomas.

(9) Tumours, Malignant

(*a*) Carcinoma (common).
(*b*) Sarcoma (rare).

APPEARANCE OF THE LARYNX

(1) Acute Laryngitis

There is usually diffuse hyperæmia. The cords may be pink, a little œdematous, or even reddened. The diffuseness of the hyperæmia is important. The mucosa remains smooth. *There may be little or no œdema. The epiglottis and pharynx often show similar* hyperæmia.

(2) Chronic Laryngitis

The mucosa is no longer smooth and flat. The hyperæmia is often not diffuse any longer. There may be ulceration or œdema, particularly in tuberculosis of the larynx.

(3) Laryngeal Paralysis

This may follow poliomyelitis, or medullary polyneuritis of unknown virus origin. Conduction along the recurrent laryngeal nerves may be interrupted by malignant disease in the cervical œsophagus, during thyroid operations, or by thyroid growths. Lesions, such as aneurism of the aorta, or inflammation at the apices of the lungs, may equally interfere with conduction along these recurrent laryngeal nerves, and cause laryngeal paralysis.

The movement of the larynx should be noted. The movement of both sides of the larynx should be symmetrical. When this normal movement is absent, the examiner

must ask himself whether some local condition such as fixation due to malignant changes, or myopathy, is preventing normal movement, or whether some lesion along the course of the recurrent laryngeal nerves is responsible. If loss of movement is caused by some local change, there will usually be signs of swelling, or obstruction, or ulceration; or there may be a past history of poliomyelitis, or diphtheria.

If the larynx looks quite normal, except for the paralysis, common sites along the courses of the recurrent laryngeal nerves worthy of clinical examination are: (1) the thyroid gland, (2) the apices of the lungs and structures related to the under surface of the aorta, and (3) the cervical œsophagus.

Absence of movement of one or both cords should be carefully noted. If one arytenoid lies a little in front of the other, this is often suggestive of unilateral paralysis, and may be helpful if the larynx is difficult to examine.

In *unilateral paralysis*, the examiner is struck by the overaction and compensation of the normal cord, which comes right across the middle line to meet the paralysed cord, if the patient endeavours to say the vowel "E." It is this remarkable *compensation* which explains the apparently normal voice in unilateral paralysis, and accounts for the tiredness of the voice towards the end of the day. There is usually no stridor, and no apparent obstruction to the airway. Compensation is rapid. Some very temporary difficulty with swallowing is experienced due to unilateral paralysis of the upper sphincter of the œsophagus (crico-pharyngeus) which is also supplied by the recurrent laryngeal nerve.

The extraordinary compensatory powers of the larynx are well seen in *bilateral paralysis*, where, although the voice is undoubtedly weak, it is remarkably clear; those muscles, not supplied by the recurrent nerves, taking on added functions. Even when the complete larynx has to be removed because of malignant changes, and a permanent tracheotomy is the only respiratory passage, many patients develop an amazingly good voice. Folds in the pharyngeal walls are used for phonation, and the relaxed œsophagus makes an efficient air reservoir.

In unilateral paralysis, the patient does not complain of shortness of breath on exertion; whereas in bilateral paralysis there is great narrowing of the laryngeal airway, because the main abductors of the cords are paralysed, and the patient does complain of real shortness of breath on exertion. There is usually acute distress if both cords are paralysed suddenly, but with reassurance, and when adaptation is established, most patients complain of little difficulty in breathing when at rest, or during light movement. The *stridor* during sleep is very inconvenient for others. There is no stridor in unilateral paralysis. It must be remembered that recovery does take place more often than is usually realized, in both unilateral and bilateral paralysis of the recurrent laryngeal nerves.

LARYNGEAL PALSIES FOLLOWING THYROIDECTOMY

Recovery depends on the lesion. If the nerve is cut right through and the ends are not in apposition, recovery will not take place. The commonest injury, however, is not due to the cutting of the nerve. It has been found during the recent war that that type of peripheral motor nerve injury which does not recover at all, although the gross continuity of the nerve appears to be intact, is due to multiple ruptures of the axons within their sheaths. If the nerve bundles are only ruptured in one place, within the perineurium,

recovery will nearly always take place. But if there are several such ruptures, recovery will not take place.

Injuries to the recurrent nerve caused by traction may be very slight, and in these cases recovery is quick and complete. In other cases, where the damage is more severe, recovery may take anything up to 2 years.

We seldom have means of finding out what particular lesion has occurred to the recurrent nerve after thyroidectomy in peace-time. Most of our information about lesions, and recovery, is based on peripheral motor nerve injuries during the war, where it was sometimes possible to dissect out the nerve concerned, and examine it under the microscope.

When very large thyroid tumours arise, the recurrent laryngeal nerves may already be stretched or displaced by the growth. Even gentle traction may cause rupture of some axons within the epineurium. If thyroid surgeons are courageous enough to tackle *all* types of growth, simple and malignant; mobile and fixed; huge and small; then occasionally they will inevitably find recurrent laryngeal paralysis as a complication to their operation. If, however, only the easiest tumours are tackled, it will not be surprising if some surgeons insist that they never have paralysis follow thyroidectomy.

Unilateral paralysis of the recurrent laryngeal nerve often passes unnoticed by patient and surgeon alike, because compensation is so good in the normal half of the larynx. All thyroid surgeons would be well advised to have the *vocal cords examined both before and after thyroid operations* for this very reason. This is particularly true if some previous operation has been performed on the thyroid, and a second operation is contemplated.

(4) Upper Œsophageal Obstruction

Froth collected in the piriform fossæ suggests upper œsophageal obstruction due to growth, foreign body, or medullary paralysis.

(5) Tumours of the Larynx

Polypi, fibromas, papillomas, hæmangeiomas, myxomas, cysts, neuromas, and all kinds of combinations (such as myxo-fibro-hæmangiomas) can occur. They leave the cords mobile, and their distinctive feature is a smooth, intact mucosa, that does not tend to bleed or ulcerate. Carcinoma, on the other hand, is distinguishable by irregular, raised epithelium, and sometimes necrotic ulceration. There is often fixation of the cords or adjacent muscles. The lesion tends to bleed easily, and the patient's breath may be offensive.

(6) Clinical Investigations

If any growth is present on either cord, the voice will never clear until the growth has been removed. Whenever the epithelium of the larynx is ulcerated, or irregular, an X-ray of the lungfields, and the Wasserman reaction, should be done before any question of biopsy is considered. If there is purulent sputum, this also should be examined.

When considering the advisability of *biopsy* as an aid to diagnosis, it must be remembered that interference with the edge of a growth (where the normal tissue cells of the body are doing their best to resist the encroachment of malignant cells) may cause rapid extension of the growth in the larynx. This is particularly important where a growth is small, and a complete cure is probable. In such cases, *the best and safest diagnostic*

biopsy is often the removal of the complete tumour surrounded by an adequate margin of healthy tissue.

No laryngoscopy is complete without an examination of the *neck*. The position of the thyroid cartilage, and trachea, should be inspected and any displacement noted. Movement of the larynx on swallowing should be free, and any unusual lump, or enlarged nodes, must be felt for.

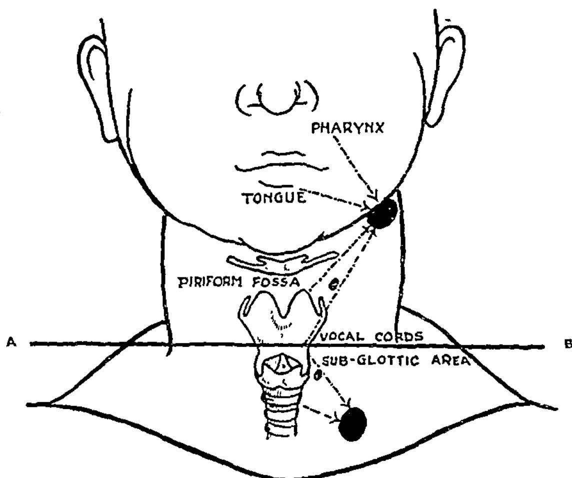


FIG. 83. *Diagram showing enlargement of lymph nodes in the neck. The line A–B is drawn through the level of the vocal cords.*

Carcinoma of the Larynx

Carcinoma, of itself, causes so little disturbance of function in many *other parts of the body*, that patients are quite unaware that they have anything wrong, until the disease is far advanced and causes obstruction, swelling, ulceration or pain. The slightest fixation or weighting of the *vocal cords*, on the other hand, causes alteration in the pitch of the voice. Not only does the patient himself notice the alteration in his voice, but his wife does too. As the voice is so necessary for the interchange of ideas within the domestic circle, and during work, hoarseness is a symptom the patient cannot keep to himself. Carcinoma of the vocal cords should, therefore, be diagnosed very early, and, if treated early, is curable with little or no deformity.

Unfortunately, carcinoma of the piriform fossæ, epiglottis and lower pharynx as a

rule do not affect the vocal cords immediately, and diagnosis is only made comparatively late in these cases.

Every effort should therefore be made by the surgeon to have cases of persisting hoarseness carefully investigated. Some general surgeons and physicians become very proficient at examining the larynx with the laryngeal mirror, but even experienced laryngologists often find the detection and description of lesions of the vocal cords very difficult, and no one should mind asking for the opinion of a laryngologist if he values the safety of his patient.

Lymph Nodes

Carcinoma of the larynx does not tend to spread to lymph nodes early. It is a well-known fact that a primary lesion of the vocal cords is diagnosed long before the lymph nodes in the neck become enlarged. The hoarseness makes early diagnosis possible. In carcinoma of the pharynx and œsophagus, however, the reverse is true. It is not uncommon to have the patient present with a lump in his neck and to discover the primary lesion in the œsophagus or pharynx only on careful examination.

Should late advanced carcinoma of the vocal cords spread to the lymph nodes of the neck, it is well for the surgeon to feel for enlargement of the nodes (1) at the angle of the jaw (superior deep cervical), and (2) over the thyro-hyoid membrane. Carcinoma below the vocal cords (in the subglottic region) tends to spread to (1) the inferior deep cervical lymph nodes, and (2) to a small node on the crico-thyroid membrane (Fig. 83).

Sexual Incidence of Carcinoma of the Larynx

Carcinoma of the larynx is much more common in men than in women. It is probable that men smoke more than women, or at any rate have been heavy smokers for longer than most women. It is also probable that men tend to come up against more air pollution in their work than do women.

TREATMENT OF CARCINOMA OF THE LARYNX: GENERAL CONSIDERATIONS

Treatment will depend on:

(1) The extent and site of the lesion.

(2) The sensitivity of the growth to irradiation.

(3) The adaptability of the patient.

Each case must be judged on its own merits, and we strongly recommend discussion between radiotherapist and surgeon beforehand, so that a course of treatment can be agreed upon that will be most likely to cure the patient of cancer, and at the same time limit deformity. Obviously the most important initial policy must be to cure the cancer. If the cancer is not destroyed, all other considerations are of secondary importance, because cancer is progressive. At the same time, the surgeon cannot concentrate only upon eradicating the cancer, because life may become unbearable if deformity and loss of function are too great a burden for the patient and his family to bear. Some patients adapt themselves to a permanent tracheotomy, and an œsophageal voice, with a courage and achievement that fills us with admiration and respect. Others find the deformity very hard to bear.

TREATMENT OF EARLY CARCINOMA, INVOLVING ONLY ONE CORD, AND WITH FREE MOVEMENT OF THE CORD

This type of case is eminently curable. It offers all the advantages that typify curable cancer elsewhere in the body. It is possible to isolate the growth entirely from the healthy living tissues with minimal trauma and no deformity. The choice lies between

(1) *Excision, by diathermy, via laryngofissure of one cord.*

(2) *Barrage by Gamma Rays from radium needles inserted beneath the length of the cord, via small holes drilled through the thyroid cartilage.*

Both methods give excellent results.

Advantages of Excision by Diathermy of Early Carcinoma of One Cord

(*a*) It is possible to send the whole excised growth to the pathologist for radial section, and so to obtain, not only a full picture of its cell structure, but also assurance that the whole growth has been completely removed (microscopically).

(*b*) Another advantage is that, by this method, the biopsy is taken, as it were, of the whole tumour, at the time of excision, and it is therefore unnecessary to cut into it beforehand.

(*c*) There is no deformity. A new cord is formed from fibrous tissue. Function of the voice should be good.

(*d*) There is very little to choose between laryngofissure and radium needles, as far as risk to the patient is concerned.

(*e*) There is no necrosis of cartilage.

(*f*) At Laryngofissure, an excellent view of the whole extent of the carcinoma is obtained.

(g) Deep X-ray Therapy can still be used after laryngofissure, if necessary.

Disadvantages of Radium Needle Barrage in Early Carcinoma of One Cord

(*a*) A biopsy has to be done beforehand, to establish malignancy, and to determine sensitivity to irradiation. This is difficult to achieve without cutting into the line of separation between carcinoma cells and healthy tissue cells. The growth is often so small that it seems a pity to cut into it, when it can be removed so easily and completely by laryngofissure.

(*b*) After both irradiation, and diathermy excision by laryngofissure, the patient has to be kept under regular observation in exactly the same way. But when irradiation is used, there is no microscopic report to reassure the surgeon that the growth has, in fact, been completely removed.

(*c*) Thickening of laryngeal cartilages sometimes takes place after irradiation, and there may be some permanent thickening and œdema of the mucosa of the larynx.

(*d*) Some malignant growths are only partially radiosensitive (papillomatous carcinomas with well differentiated cells).

Advantages of Radium Needles in Very Early Carcinoma of One Cord

(*a*) Irradiation can seek out and destroy isolated spreading malignant cells, particularly if the growth edges are not clearly defined.

(*b*) There is no deformity. Function of the voice should be good.

(c) The larynx does not have to be opened. Both methods, however, require considerable skill.

Other methods for destruction of early carcinoma of one cord are:

(1) **Removal with cup forceps from above via a laryngoscope.** It is so difficult to be sure that the whole growth is removed by this method. One cannot see beneath the cord. We have discarded this method now that we have perfected laryngofissure. We say this, even though we have patients still alive and well, and with normal voice, 8 years after removal of proved carcinoma by this method.

(2) **Diffuse Irradiation by Deep X-ray Therapy.** This method gives very good results. The great advantage is that no surgical operation whatever has to be carried out. But Deep X-ray Therapy is not as innocuous as having one's photograph taken. The patient sometimes has a fairly unpleasant time; there may be telangiectatic changes and discoloration of the skin; there may be irradiation thickening, or even necrosis, of the laryngeal cartilages. Of course, in the hands of a careful and skilled radiotherapist, the risk of these disadvantages is minimal.

As stated above, the only wise procedure is for radiotherapist and surgeon to discuss therapy and technique together beforehand.

OPERATION OF LARYNGOFISSURE

This operation, as performed at the London Hospital, was the outcome of close co-operation between surgeon and anæsthetist to seek a technique that would avoid tracheotomy. The author is greatly indebted to Dr. A. I. Parry Brown for his great skill, and loyal partnership, in contributing so very much to the success of this procedure. The technique employs the principle of operating round the endotracheal tube, after incising the thyroid cartilage in the midline, and separating the two vocal cords without disturbing their attachments.

We first performed this operation in 1941 and have had ample experience since. The technique has everything to recommend it. Recovery is rapid. The patient speaks immediately on return to consciousness. There have so far been no complications, except a variable degree of surgical emphysema, in some cases, which has given no trouble. There is no deformity. The scar need be no more visible than a good thyroidectomy scar, and not nearly so long Complete removal of the growth is made possible using magnification $\times$ 2. Hæmostasis is assured.

This operation is not only used for very early cases of carcinoma of the vocal cords, but for *removing non-malignant tumours of the cord* where complete accurate removal by endoscopy is considered to be unlikely.

ANÆSTHESIA FOR LARYNGOFISSURE

There are three requirements—first, a quiet surgical field free from reflex activity. Second, the anæsthetic tube must not interfere with surgical access. Third, the bronchi must be protected from any blood spilt during the operation.

Our technique is to obtain local analgesia of the larynx by the injection of 1 ml. of 10 per cent cocaine HCl through the crico-thyroid membrane. The induction of anæsthesia is by injection of thiopentone, about twice the sleep dose. At this stage, the jaw is usually relaxed and intubation under direct vision is simple. If any difficulty is experienced then a small dose of short acting relaxant will facilitate the intubation. The

tube chosen should be about a No. 5 Magill, as any larger tube will obstruct the view of the cords when the larynx is opened.

Its length should be judged to bring its tip within 1 cm. of the carina, as packing must be introduced into the trachea below the laryngofissure in order to isolate the surgical field from the bronchial tree. The maintenance of anæsthesia is by nitrous oxide, oxygen, and chloroform. This mixture gives quiet breathing, little bleeding and freedom from reflex activity.

During the course of the operation, the anæsthesia is deepened immediately before the opening of the larynx, but the chloroform can be withdrawn as soon as the pack has been removed and the larynx closed.

At the end of the operation the tube is removed under direct vision in order that any blood can be aspirated from the pharynx.

Position on the Table

Head extended over a soft pillow 5–10 degrees.

The extended position of the head and neck brings the larynx and trachea up out of the thorax. In this position the lower border of the cricoid may lie as much as 8 cm. above the suprasternal notch in a young male adult, but quite often the length of the trachea above the suprasternal notch may be very much less.

Towels are placed in position.

The neck is palpated with the fingers, holding the hand flat. The larynx and trachea should be gently moved over the bodies of the cervical vertebræ until the ring of the cricoid cartilage can be clearly identified. The trachea is compressible, being composed of horse-shoe like rings, but the firm inflexible cricoid gives the surgeon his exact position. This is very important. In some patients with a short fat neck, and particularly in women, it may be extremely difficult to identify even the thyroid cartilage. A transverse incision is made through the skin of the neck, near the lower border of the thyroid cartilage, about 2½ in. long. The incision is made through the skin, and platysma, with the knife at an angle, so that, when the skin comes together after the operation, the upper lip will lie on the lower lip, rather like a skin-graft (this oblique cutting gives a much less visible scar).

The skin flap is dissected upwards to give just sufficient exposure of the thyroid cartilage. There are many advantages about a vertical incision, but the transverse one probably leaves a less conspicuous scar.

The whole idea of the operation is to disturb the tissues of the neck as little as possible.

The anterior jugular veins are carefully dissected, so that they can be retracted laterally, and need not be divided. Tissue planes are disturbed as little as possible. The thyroid cartilage is exposed by separating the sterno-hyoid muscles in the midline, or by dividing the fibres of one longitudinally. If this is done, the muscle fibres can be brought together after the operation to seal off the larynx more securely, and there is less likelihood of surgical emphysema.

The sternohyoid muscles are retracted laterally and the full length of the thyroid cartilage is now visible. The perichondrium of the thyroid cartilage is divided in the midline, but only a thin strip in the midline is bared of perichondrium. Denuding the cartilage predisposes to necrosis.

The cricothyroid ligament is cleaned. The point where the anæsthetist has injected

his subglottic analgesic, is seen in the centre of this ligament. The length of the ligament varies greatly from patient to patient. A small artery, a branch of the superior thyroid, runs across the ligament and invariably enters the larynx through a perforation in the centre of the ligament. This small artery should be nipped with artery forceps and clotted before opening the larynx. In some young adults, the thyroid cartilage is soft and elastic, and can be cut through with the knife in the midline. In most adults, however, ossification is advanced, and an electric drill is used to cut through the midline down to, but not entering, the mucosa. The surgeon must remember that the vocal cords are attached to the thyroid cartilage within, on either side of the midline, by clearly defined separate insertions. The object of the next step is to cut dead between their respective insertions, and so avoid displacement. The second anatomical point to remember is that these attachments of the cords to the inner surface of the thyroid cartilage lie at a point between the upper two-thirds, and lower third, of the anterior border of the thyroid cartilage. The vocal cords lie in their entirety *above* the cricoid cartilage, being attached posteriorly to the vocal processes of the arytenoid cartilages; which cartilages rest upon the upper surface of the posterior (signet ring) aspect of the cricoid.

All should now be made ready for entering the larynx. Strips of continuous gauze, attached to artery forceps to prevent loss, are prepared to pack off the trachea below, as soon as the larynx is opened. The anæsthetist is asked for permission to open the larynx (this is important) and an incision is made through the centre of the cricothyroid ligament up through the slit in the thyroid cartilage, between the insertions of the cords. A special retractor is placed in position. The two halves of the thyroid cartilage are gently separated. Gauze is carefully and lightly packed down the sides of the trachea, round the endotracheal tube. The growth is inspected and dissected out. The endotracheal tube gives little inconvenience. It is easily pressed gently to one side to facilitate accurate dissection if necessary.

Magnification. Magnification × 2·5, and a headlamp, are used for inspection, and during dissection.

Diathermy. For dissection, coagulation, 2, throughout.

Suction. For removal of mucus or blood from the larynx, but should not be allowed to disturb fibrin.

Hæmostasis. The edges of the mucosa are sutured together with interrupted atraumatic fine catgut sutures, using an ophthalmic needleholder. The self-retaining retractor is removed, and the gauze packing is removed. The cricothyroid membrane and the thyroid perichondrium are sutured, and the wound closed in layers. The skin wound is closed with interrupted waxed-thread mattress sutures. A dry gauze dressing is applied. We do not use a drain, but one could be left in, just under the skin, if hæmostasis is not perfect.

Post-operatively the patient is propped up in bed as soon as consciousness is fully regained. Speaking is encouraged, but excessive coughing, straining, and sneezing are discouraged as far as possible, to minimize surgical emphysema. Swallowing gives little or no difficulty. Skin sutures are removed on the fifth day.

Treatment of Invading or Advanced Carcinoma of the Larynx

(1) If both vocal cords are involved, or there is extension of growth to the anterior commissure, or invasion of the ventricle, false cord, arytenoid eminence, or trachea, we

think radiotherapy is indicated—usually Deep X-ray Therapy, unless the spread is well localized.

(2) Some surgeons would advocate laryngectomy in these cases, as a first procedure, with a good chance of complete cure, at the price of a permanent tracheotomy and all that that means.

(3) It is a difficult decision to make. One has to consider each individual case. The kindest thing is to try to think what one would want done for oneself, or one's closest relatives; remembering that the patient may not altogether feel as we do about these things.

(4) Whilst admitting that early laryngectomy will give a very good chance of total eradication of this type of carcinoma, we favour radiotherapy first. A good radiotherapist will soon know whether the growth is responding, and will advise laryngectomy in plenty of time to save the patient.

(5) There are the usual disadvantages of irradiation—thickening of the laryngeal cartilages, sometimes laryngeal obstruction, or laryngeal necrosis. Tracheotomy may have to be performed. So much depends upon the extent of the growth and the dosage and application of radiotherapy.

(6) We have seen excellent results from Deep X-ray Therapy to spreading carcinoma of the larynx, and feel we must give our radiotherapy colleagues a chance, when we know they can cure our patients without sacrificing the larynx.

(7) Laryngectomy can always be performed if radiotherapy fails.

ANÆSTHESIA FOR LARYNGECTOMY

The first stages of a laryngectomy are conveniently carried out with the anæsthetic technique described for laryngofissure, but the anæsthetic tube must be withdrawn when the trachea is divided. A short, cuffed, endotracheal tube with its connections is sterilized and is inserted into the trachea by the surgeon. The cuff is inflated and careful observation made that both lungs are inflating to ensure that the end of the tube is not endobronchial.

Maintenance of anæsthesia through this tube presents no special difficulties, but two points are worthy of mention. If there has been long-standing respiratory obstruction, the respiratory centre may be responding only to a raised tension of CO_2 in the blood. The relief of obstruction by tracheotomy may be followed by respiratory depression. It is wise to avoid respiratory depressants in the pre-medication.

If the operation is so extensive that it is expected to take more than 2 hours, it is inadvisable to use chloroform because of its toxic action. Trilene will usually provide sufficient depth of anæsthesia once the tracheotomy has been performed.

Position on the Table

The patient will lie on his back with his head extended over a soft pillow 5–10 degrees.

A U-shaped incision is made from the level of the hyoid 2 in. from the midline to the supraclavicular fossa. The skin-and-platysma flap is dissected upwards, and bleeding points are coagulated with diathermy. The pre-tracheal muscles are separated in the midline and retracted laterally. The isthmus of the thyroid is separated from the trachea and divided between clamps. Hæmostatic sutures are inserted into the cut ends.

The trachea, cricoid, and thyroid cartilages and the hyoid bone are dissected from the

overlying fascia. The sternothyroid and thyro-hyoid muscles are separated from the thyroid cartilage by blunt dissection. This dissection is continued laterally to separate the inferior constrictor muscle from the cricoid and thyroid cartilages. At this stage the inferior laryngeal artery is identified at the lower border of the thyroid; it is ligated and divided. The recurrent laryngeal nerve is found with the artery and is divided.

The thyroid cartilage is freed above by dividing the upper cornu as high as possible. The trachea is divided at about the level of the second or third ring, according to the spread of the growth, and dissecting upwards from the anterior œsophageal wall. This dissection must be done with considerable care, as the posterior wall of the trachea, which is made up of soft elastic muscle and not cartilage, has to be separated from the relatively thin anterior wall of the œsophagus. The separation is continued upwards and the longitudinal œsophageal muscles are separated from their insertion into the posterior plate of the cricoid. When the trachea is divided, the endotracheal anæsthetic tube is withdrawn and replaced by a new, short, cuffed tube, which is inserted into the lower end of the trachea.

Dissection is continued upwards posteriorly, so that the whole larynx, up to the tips of the arytenoids, is dissected from the anterior œsophageal wall. The pharynx is then entered, and the mucosa of each piriform fossa is carefully separated from the larynx and preserved. The thyro-hyoid membrane is next divided and the epiglottis is dissected from the base of the tongue. A Ryle's tube is passed at this stage into the stomach. Its upper end is passed up into the mouth, whence it is brought up behind the soft palate and through the nostril. Its end is secured to the cheek with a small strip of adhesive.

The larynx and upper part of the trachea are now free and are removed. They are inspected carefully for any suspicious areas that may suggest invasion of the œsophageal wall or piriform fossæ. If any suspicious area is found, the neighbouring tissue in the pharynx or œsophagus is excised and sent for microscopic examination. The mucosa of the piriform fossæ is used to close off the pharynx, and is sutured with interrupted black waxed threads. The pre-tracheal muscles, which have been preserved, are sutured over this suture area to give extra strength to the anterior œsophageal wall and to help close off the pharynx. The skin-flap is sewn down to the upper end of the posterior wall of the trachea No stretching should be permitted. The rest of the skin-flap is sutured to the sides of the trachea and to the sterno-mastoid muscle sheaths. Bare areas are thus left on either side still exposing muscle. These areas are skin-grafted from the thigh. This technique gives a good, strong, mobile anterior œsophageal wall. The extent of the operation must vary according to the invasion of the growth, and parts of the pharyngeal wall, tongue, hyoid, or œsophagus may have to be included. If much œsophageal wall has to be sacrificed, the œsophagus may have to be reconstructed, and skin-grafting can help to close the gaps. Broadly speaking, laryngectomy gives excellent results, provided the growth is contained within the thyroid cartilages and larynx. If spread has taken place to the tongue, or if it involves the œsophagus or pharynx extensively, laryngectomy will often fail to cure. In all cases, discussion with the radiotherapist should take place both before and after laryngectomy.

Post-operative treatment will continue along the lines laid down for tracheotomy. The patient writes on a pad his nursing requests and answers. For the first few days the Ryle's tube is used for feeding, but the patient can be encouraged to swallow water and fluids if no extensive reconstruction of the pharynx or œsophagus has had to be

undertaken. After healing and adjustment to the tracheotomy, instruction in œsophageal speaking must be given by a speech therapist.

The Œsophageal Voice after Laryngectomy and Permanent Tracheotomy

Normally we speak by expelling air from our lungs through the vocal cords, pharynx, and mouth. The cords give us pitch, our tongue and pharynx give us quality. No air enters the œsophagus during respiration, or during actual speech, because the upper sphincter (crico-pharyngeus) keeps the œsophagus closed. We cannot even swallow during actual respiration or actual speech.

When laryngectomy is performed, the upper sphincter of the œsophagus is detached from the cricoid cartilage (its only fixed insertion) and cannot keep the œsophagus closed efficiently. On deep inspiration negative pressure in the chest opens the œsophagus and enables the patient to draw air into the œsophagus. This, in spite of the fact that the only entrance to the patient's lungs is now through the tracheotomy opening in the neck. There is, of course, no communication whatsoever between the patient's lungs and the patient's pharynx, nose, or mouth, after laryngectomy. Thus air which enters the œsophagus on deep inspiration can be used for speech. The air does not enter the stomach because the lower sphincter remains closed at the cardia. The patient learns to expel this œsophageal air through his pharynx, and so is able to speak. He has no larynx, but nature is so incredibly adaptable in this area that folds in the muscles and connective tissue of the lower pharynx act as an artificial larynx.

Some patients speak most efficiently, others seem to have great difficulty. One cannot help feeling, after observing a number of these patients, that success depends more on the patient, and his ability to help himself, than it does on the speech therapist. But the speech therapist can explain to him what to do. He does the rest.

When major excisions in the neck are contemplated, such as laryngectomy with block dissection of glands of neck, or laryngectomy, with partial removal of other structures variously invaded by malignant disease (pharynx, œsophagus, tongue, thyroid), it is of enormous help to *work with another experienced surgeon.* The author of this article owes much to his general surgical colleague, Mr. Clive Butler, for his kind and sympathetic co-operation in difficult undertakings. This team work is to be recommended highly to those who may not as yet practise it. It requires mutual trust and a certain humility, irrespective of seniority.

In difficult neck cases, fine judgment is often required *after* a long and hazardous dissection. It is of enormous benefit to the patient at such times if the surgeon in control can discuss the situation with an experienced colleague on the spot, or hand over completely to him. Mr. Butler, and the author of this short article, slip in and out of their rôles, as surgeon in complete charge, and first assistant, with the greatest of ease, and without the slightest reservation. This happy combination of general surgeon and specialized surgeon in big dissections of the neck, mouth, or pharynx, has much to recommend it. It cannot but impress students and nurses alike with its sense of sublimation to the main purpose of the operation—the cure of the unconscious patient.

LARYNGEAL STRIDOR IN INFANTS

(1) A common cause is *subluxation of one arytenoid eminence* into the laryngeal aperture on deep inspiration. There is stridor on deep inspiration. This condition occurs

overlying fascia. The sternothyroid and thyro-hyoid muscles are separated from the thyroid cartilage by blunt dissection. This dissection is continued laterally to separate the inferior constrictor muscle from the cricoid and thyroid cartilages. At this stage the inferior laryngeal artery is identified at the lower border of the thyroid; it is ligated and divided. The recurrent laryngeal nerve is found with the artery and is divided.

The thyroid cartilage is freed above by dividing the upper cornu as high as possible. The trachea is divided at about the level of the second or third ring, according to the spread of the growth, and dissecting upwards from the anterior œsophageal wall. This dissection must be done with considerable care, as the posterior wall of the trachea, which is made up of soft elastic muscle and not cartilage, has to be separated from the relatively thin anterior wall of the œsophagus. The separation is continued upwards and the longitudinal œsophageal muscles are separated from their insertion into the posterior plate of the cricoid. When the trachea is divided, the endotracheal anæsthetic tube is withdrawn and replaced by a new, short, cuffed tube, which is inserted into the lower end of the trachea.

Dissection is continued upwards posteriorly, so that the whole larynx, up to the tips of the arytenoids, is dissected from the anterior œsophageal wall. The pharynx is then entered, and the mucosa of each piriform fossa is carefully separated from the larynx and preserved. The thyro-hyoid membrane is next divided and the epiglottis is dissected from the base of the tongue. A Ryle's tube is passed at this stage into the stomach. Its upper end is passed up into the mouth, whence it is brought up behind the soft palate and through the nostril. Its end is secured to the cheek with a small strip of adhesive.

The larynx and upper part of the trachea are now free and are removed. They are inspected carefully for any suspicious areas that may suggest invasion of the œsophageal wall or piriform fossæ. If any suspicious area is found, the neighbouring tissue in the pharynx or œsophagus is excised and sent for microscopic examination. The mucosa of the piriform fossæ is used to close off the pharynx, and is sutured with interrupted black waxed threads. The pre-tracheal muscles, which have been preserved, are sutured over this suture area to give extra strength to the anterior œsophageal wall and to help close off the pharynx. The skin-flap is sewn down to the upper end of the posterior wall of the trachea. No stretching should be permitted. The rest of the skin-flap is sutured to the sides of the trachea and to the sterno-mastoid muscle sheaths. Bare areas are thus left on either side still exposing muscle. These areas are skin-grafted from the thigh. This technique gives a good, strong, mobile anterior œsophageal wall. The extent of the operation must vary according to the invasion of the growth, and parts of the pharyngeal wall, tongue, hyoid, or œsophagus may have to be included. If much œsophageal wall has to be sacrificed, the œsophagus may have to be reconstructed, and skin-grafting can help to close the gaps. Broadly speaking, laryngectomy gives excellent results, provided the growth is contained within the thyroid cartilages and larynx. If spread has taken place to the tongue, or if it involves the œsophagus or pharynx extensively, laryngectomy will often fail to cure. In all cases, discussion with the radiotherapist should take place both before and after laryngectomy.

Post-operative treatment will continue along the lines laid down for tracheotomy. The patient writes on a pad his nursing requests and answers. For the first few days the Ryle's tube is used for feeding, but the patient can be encouraged to swallow water and fluids if no extensive reconstruction of the pharynx or œsophagus has had to be

demonstrate inclusion bodies or viruses in cases coming under our care. The immediate risk is laryngeal obstruction, and the immediate consideration may be tracheotomy. The late risk is stenosis of the larynx due to scarring and permanent loss of voice with permanent tracheotomy. Destruction of the growths by diathermy we think is unwise, because intense heat is a common cause of laryngeal stenosis. We prefer removal by cup-cutting forceps, via a laryngoscope, under some such anæsthetic as rectal avertin. The papillomata of *only one side* of the larynx are removed. The operation is repeated for the other side when healing has taken place. In this way the voice will be preserved, and stenosis and tracheotomy avoided. Care must be taken not to injure the cord, or underlying muscle, when removing the papillomata. The patient is kept under periodic observation, and laryngeal obstruction is prevented by repetition of this procedure. The growths do not tend to recur when puberty approaches. We have proved the efficacy of this management from infancy to puberty, and, until such time in the future when papillomata can be treated otherwise than by removal, we find it very satisfactory. An alternative method is to treat the papillomata by radiotherapy, but the long-term effects of irradiation on a child's larynx have to be considered.

TRACHEOTOMY

The main difficulty about this operation is to know *when to do it.* A successful tracheotomy short circuits respiration by allowing the patient to breathe through the trachea direct, thus by-passing the nasal passages, the pharynx, and the larynx.

As far as possible, tracheotomy should be performed *before it becomes immediately urgent.* It can then be done with minimal risk to the patient. If, therefore, respiratory embarrassment is becoming worse in a patient with laryngeal œdema, and the obstruction is likely to continue, or even become more complete, tracheotomy should not be postponed too long. This is especially important in spinal and medullary paralysis from polioencephalitis or virus-polyneuritis, when intermittent positive pressure respiration may have to be carried out for several days or weeks. Such patients will be partially paralysed in any case, and the paralysis will probably be progressive. The intention in these cases, is to connect the tracheotomy tube with an automatic breathing machine.

Again, when progressive carcinoma of the larynx or pharynx is causing severe respiratory obstruction, or when treatment by radiotherapy is about to be undertaken in a patient with considerable respiratory obstruction, it is wiser to perform tracheotomy before it becomes really urgent. There is no sense in waiting until cerebral anoxia has permanently injured the brain.

The second point is to realize that it is very seldom necessary today to open the trachea in a semi-asphyxiated and congested patient. Modern anæsthetic practice has given us the *endotracheal tube*, and in most cases of paralysis or laryngeal obstruction, it is possible to pass an endotracheal tube as a preliminary before tracheotomy. The tube is passed under direct vision, and immediately establishes an airway. This relieves venous congestion and prevents cerebral anoxia, and gives the surgeon time to prepare for tracheotomy. The tracheotomy can then be performed unhurriedly, as it were, over the endotracheal tube; the latter being withdrawn slowly as the tracheotomy tube is inserted. This has so many advantages over a hurried operation on a critically congested patient, that it should always be tried whenever possible.

in new-born infants. It is readily seen on direct laryngoscopy, and most children lose their stridor as laryngeal muscles compensate for joint weakness. The lesion may possibly be traumatic, or may be due to myopathy of virus origin.

(2) *Paralysis of one vocal cord* occurs in infants with congenital enlargement of the left side of the heart.

(3) *Congenital webs* of the larynx. The larynx, trachea, and bronchi are developed from a ventral fold in the wall of the œsophagus (fore gut), and this knowledge helps the surgeon to understand the position of such deformities. They may give little embarrassment to respiration, but sometimes require immediate attention, when the surgeon is faced with two alternatives—tracheotomy or removal of the web. The latter is obviously the more rational and curative therapy, because there is usually nothing else wrong with the child. Some infants will not survive tracheotomy. Both alternatives carry a considerable element of risk, and respiratory embarrassment and cerebral anoxia may force the surgeon's hand. There is stridor both on inspiration and expiration.

(4) *Cysts of the Larynx*. These are not common, but if recognized are comparatively easy to treat, and successful treatment will save useful lives because there is usually nothing else wrong with the patient. The clinical picture is that of an infant with respiratory obstruction at the larynx, and hypersecretion of pharyngeal mucus. There is often severe stridor on inspiration. These babies tend to drown in their own pharyngeal secretions because the cyst may cause partial obstruction to the œsophageal opening. The cyst, often large, and lying laterally, can be aspirated with a long needle using a laryngoscope, and anoxia is immediately relieved. This will save life. Later the cyst can be dealt with by excision.

(5) *Hæmangiomata* of the neck produce laryngeal obstruction and stridor. The infant usually gives no indication of obstruction at all until, during a feed, a little milk inadvertently enters the laryngeal inlet. In a normal infant this causes distress, spasm, and cyanosis, from which recovery is rapid. If diffuse hæmangiomatous changes are present in the infant's neck (and there may be only slight patchy skin changes over widely distributed hæmangiomatous changes in the deep tissues of the neck), the blood spaces become engorged and the laryngeal obstruction may become so progressive, that tracheotomy becomes imperative, even though the surgeon realizes all the later implications of this procedure. It may be very difficult to do away with the tracheotomy opening.

(6) *Spasmodic Laryngitis*. This is a spasmodic contraction of the laryngeal sphincter muscles, leading to closure of the glottis, and progressive dyspnœa. Convulsions may occur. Recovery is usual. The possible cause may be irritation from adherent mucus on the cords during sleep, because the spasm often occurs on waking. In small children it may be a purely nervous affection without any inflammatory condition of the larynx. It is common in rickets, and in children with hyperplasia of adenoid tissue in the nasopharynx; in both of which conditions catarrhal changes predispose to adherent mucus in the larynx. The spasm may be set off by entrance of mucus or saliva into the larynx in infants waking from sleep after a period of mouth breathing.

Multiple Papillomata of the larynx in young children can be very distressing. Diffuse warty-like growth processes may replace most of the normal epithelium of the laryngeal inlet, causing severe respiratory embarrassment. The epithelium is intact over the growth processes, and bleeding is not common. These growths are thought to be of virus origin, but in spite of careful search by eminent workers on viruses, we have not been able to

demonstrate inclusion bodies or viruses in cases coming under our care. The immediate risk is laryngeal obstruction, and the immediate consideration may be tracheotomy. The late risk is stenosis of the larynx due to scarring and permanent loss of voice with permanent tracheotomy. Destruction of the growths by diathermy we think is unwise, because intense heat is a common cause of laryngeal stenosis. We prefer removal by cup-cutting forceps, via a laryngoscope, under some such anæsthetic as rectal avertin. The papillomata of *only one side* of the larynx are removed. The operation is repeated for the other side when healing has taken place. In this way the voice will be preserved, and stenosis and tracheotomy avoided. Care must be taken not to injure the cord, or underlying muscle, when removing the papillomata. The patient is kept under periodic observation, and laryngeal obstruction is prevented by repetition of this procedure. The growths do not tend to recur when puberty approaches. We have proved the efficacy of this management from infancy to puberty, and, until such time in the future when papillomata can be treated otherwise than by removal, we find it very satisfactory. An alternative method is to treat the papillomata by radiotherapy, but the long-term effects of irradiation on a child's larynx have to be considered.

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Some cases of severe obstruction, from carcinoma or foreign bodies, render the preliminary passage of an endotracheal tube impossible or unwise. This can easily be determined when the larynx is inspected with the laryngoscope prior to the passage of the tube.

In the days when diphtheria was very prevalent and the disease caused many deaths in young children, laryngeal diphtheria was quite common and caused obstruction. Tracheotomy was a common life-saving operation. It had to be performed, as often as not, under most unfavourable, and even dangerous, conditions. Any general practitioner might have found himself being forced to perform the operation to save life. It was quite common practice in fever hospitals. Today, immunization of young children against diphtheria has almost wiped out the disease. The large collections of tracheotomy sets in fever hospitals are still silent evidence of those days of not so long ago, and another reminder to the surgeon of today of the great advances of preventive medicine. It is interesting to note that even before rubber endotracheal anæsthetic tubes were invented, intubation for immediate relief was performed by passing a short metal tube into the larynx. The tubes had a lip or flange, something like a Souttar's tube, but they were not flexible. The lip prevented the tube from slipping down the trachea into the bronchi. The tube was passed over the back of the tongue with a metal introducer, and had threads attached so that it could be retrieved later on. These too can be seen in fever hospitals today.

It will always be argued that the passage of an endotracheal tube may dislodge fringes of diphtheritic membrane or friable carcinoma. This is true and the risk must be considered. It might indeed be foolish to pass an endotracheal tube if bleeding was likely to be profuse. Under those circumstances it might probably be wiser to perform tracheotomy without previous intubation. It must be remembered, however, that with modern suction apparatus most of these objections can be overcome. Anyone who has really been forced to perform hurried tracheotomy on a severely asphyxiated patient will appreciate the enormous advantages of reducing anoxia and congestion beforehand.

Surgeons are still forced to perform tracheotomy as an emergency, and without preliminary intubation. Indeed, it might still fall to the lot of any doctor to do this operation. A young patient, with a retracted lower jaw, scarred from an old osteomyelitis, became choked with teeth impacted in his larynx, during nitrous oxide anæsthesia. The writer was working in another department. It was a matter of seconds almost; there was no time even to wash.

DISADVANTAGES OF TRACHEOTOMY

(*a*) A patient with a tracheotomy tube in his trachea can no longer cough up mucus from his lungs efficiently.

(*b*) He can no longer avail himself of the air-conditioning physiology of his upper respiratory tract, which normally:

(i) Filters dust particles and bacteria from the inspired air (cilia and mucus).
(ii) Warms the inspired air (arterio-venous blood spaces).
(iii) Moistens it (profuse mucus-secreting glands).

(*c*) He can no longer speak properly, because no air is now passing through his larynx.

(*d*) He can no longer lift things as efficiently as he could before, because he cannot fix his chest by closing his larynx.

(*e*) He can no longer smell properly, because air cannot be drawn up to his olfactory receptors efficiently.

He will therefore need, in the early stages:

(*a*) Constant attention. His fears and anxieties must be allayed by good nursing.

(*b*) Removal of mucus by intermittent suction. At first he will require this at frequent intervals. If purulent bronchitis is already present, this will be doubly necessary.

(*c*) Protection from dust; a gauze apron is hung over the tube.

(*d*) Protection from cold air; open windows, early morning draughts.

(*e*) Artificial humidity if necessary with a half-steam-tent.

(*f*) The nursing staff should be ready to lift him when necessary to save over-exertion.

(*g*) He should be protected from escaping gas, or smoke, because he cannot protect himself.

Compensation will gradually adjust itself, and in time, a well performed tracheotomy gives very little trouble; indeed, it may pass unnoticed. At first there will be difficulties. If there is no infection, the worst will be over in a week or two. So much depends upon whether infection is present, and on the cause of the laryngeal obstruction. A patient with necrosis of the larynx will not have a happy time.

When tracheotomy is performed because of respiratory paralysis, the tracheotomy tube is fitted with an inflatable cuff, so that there is no leak between the tube and the tracheal walls. The tracheotomy tube will be connected with a portable automatic breathing machine. Incorporated in this machine will be a thermostat, a humidifier, and a filter. Suction of mucus from the trachea will be applied intermittently.

There are, of course, risks to the patient associated with the actual performance of the operation.

(*a*) Bleeding. This may be profuse, from the anterior jugular veins, if there is severe congestion, and inspired blood will not help a patient paralysed with poliomyelitis.

(*b*) Infection. The less the connective tissue and fascia is disturbed at operation the better. Mucus will contaminate all tracheotomy wounds. Inflammation may supervene. Necrosis of tracheal cartilages does very occasionally follow.

(*c*) Inability to relieve obstruction. No relief will be gained from a tracheotomy if the obstruction persists below the tracheotomy opening. This is important to remember in carcinoma of the bronchus or thyroid gland.

ADVANTAGES OF TRACHEOTOMY

(1) If the respiratory tract is severely obstructed above the trachea, and cannot be relieved, tracheotomy can save life by restoring the airway in asphyxia.

(2) In spinal and medullary paralysis from polioencephalitis or other cause, tracheotomy enables intermittent positive pressure respiration to be maintained for long periods.

(3) Tracheotomy can relieve respiratory distress, and so permit treatment, such as radiotherapy, to malignant disease involving the larynx.

(4) Tracheotomy occasionally affords a temporary respiratory tract, and an anæsthetic channel, when some severe surgical operation, that would temporarily obstruct

the airway, has to be performed, and which makes the use of a long nasal or oral endotracheal anæsthetic tube impracticable.

Operation

Like appendicectomy, tracheotomy can be a relatively simple operation. Certainly it is an operation that every general surgeon should be prepared to perform. It can, however, be a really formidable undertaking if there is (*a*) asphyxia, (*b*) carcinoma of the thyroid, (*c*) scarring from an old thyroidectomy.

The term tracheotomy is used because it is the familiar term in general use. Tracheostomy might be the better word for this operation of short circuiting respiration for anything except momentary relief.

The operation of tracheotomy used to imply the simple opening of the trachea, and this was performed by cutting through the anterior wall of the trachea with a vertical incision. Today, it is more usual to cut out a small round opening to fit the tracheotomy tube. This is much more satisfactory than a longitudinal slit if the tracheotomy tube is to be left in position for some length of time, and particularly if intermittent positive pressure respiration is to be carried out.

Position. An endotracheal tube is first passed under direct vision, past the obstruction. Cyanosis is relieved. The patient should be placed lying on his back on the operating table, with the head extended over a soft pillow, which is placed under the shoulders and supports the neck. After cleansing the skin, the surgeon should inspect his operation field. The notch made by the manubrium sterni is always visible and palpable. On each side of the notch, are the folds made by the anterior margins of the sternomastoid muscles. In most patients they are clearly seen, but in plump infants, and patients with fat, rounded necks, they have to be felt for. They are very easily seen in an elderly patient with carcinoma. Above, in the middle line, is the prominence of the thyroid cartilage or "Adam's Apple." In some patients this is easily identified, but in patients with obtuse angled thyroid cartilages, and plump necks, this landmark, too, can be very difficult to identify.

The surgeon should solve his difficulties by placing the flat of his hand on the front of the neck, and should gently roll the larynx and trachea over the bodies of the cervical vertebræ. In the extended position, the cricoid cartilage is easily identified in all patients. It is felt as a broad ring under the fingers, because it is the *only* cartilage that is complete. All the other bones and cartilages of the pharynx and larynx are incomplete behind (hyoid, thyroid, tracheal cartilages). Only the cricoid will resist gentle pressure with the flat of the hand. Towels are adjusted.

Incision. A horizontal incision is tempting because it lies in the fold of the neck, and such incisions for thyroidectomy often leave almost invisible scars. It must be remembered, however, that tracheotomy, unlike thyroidectomy, will communicate with the trachea, and the incision will inevitably become contaminated with mucus, which may even be purulent if bronchitis is present. A vertical incision will disturb less anatomy, and in an acute emergency, is the incision of choice.

The incision should be long enough to enable the surgeon to see what he is doing, and short enough to limit disturbance of neck structures. With the fingers on the cricoid ring, the incision passes over the ring and downwards for about 2½ in. in the middle line. Depending on the circumstances, the incision may be longer, and can be shorter. If a

horizontal incision is to be used, the incision should lie about ½ in. below the cricoid. Skin and platysma are reflected up, and the pretracheal (sterno-hyoid) muscles are seen with the anterior jugular veins running longitudinally up and down on either side of the middle line. They can look quite formidably large sometimes, and they vary a little. These veins are the source of all the distressing bleeding sometimes encountered, and a quick study of their positions will enable the surgeon to avoid injuring them except where necessary (Fig. 84).

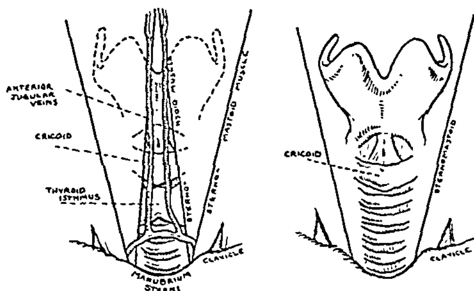


FIG. 84 Applied anatomy for tracheotomy.

They run down from the hyoid bone, close to the middle line, to about the level of the third or fourth tracheal ring, where they join one another across the middle line; then they pass under the edge of the sternomastoid, on each side, to empty into the termination of the external jugular vein, or into the subclavian vein.

The cricoid ring, which is often the only structure identifiable with certainty, is exposed by separating the two sterno-hyoid muscles, and the two anterior jugular veins. The incision is carried down through the pre-tracheal fascia. The pretracheal muscles and veins are retracted laterally. By blunt dissection, keeping close to the trachea, the soft tissues are separated from the trachea from above downwards. This can be done with the finger in an emergency. Two small artery forceps are introduced, from the exposed cricoid downwards; with the deep blade lying on the tracheal rings, and the superficial blade lying above the soft tissues. They lie side by side, and clamp the pre-tracheal junction between the external jugular veins, and the isthmus of the thyroid gland. The tissues are cut between the two clamped forceps, which are gently retracted, and the trachea is now fully exposed with no bleeding.

The *thyroidea ima* artery never gives trouble if this technique is adopted. A large *pyramidal lobe* of the thyroid can puzzle the surgeon unless he remembers his embryology. It is easily retracted laterally. *Scarring*, or inflammation can make the approach extremely difficult, because it confuses the anatomy, and the presence of a malignant growth can make the dissection formidable.

In some patients the *trachea* appears to dip down in a disheartening way on its passage

into the thorax, particularly if the cervical vertebræ are arthritic and the neck is fixed in a forward position (kyphosis).

Length of trachea exposed. In some patients the trachea seems quite long, in others, distressingly short. The surgeon must remember that the trachea rises out of the thorax, and returns to the thorax, over a wide range of movement. In a young adult, in the extended position, the length of trachea between the lower border of the cricoid and the suprasternal notch will be 8 cm. Yet, if the chin is dropped on the sternum in complete flexion, even the thyroid cartilage enters the thorax. Scarring round the thoracic inlet, substernal goitres, or fixation by growth, can limit greatly the movement of the trachea out of the thorax.

If, because of obstruction below the cricoid involving the upper trachea, low tracheotomy must be performed, the same technique is adopted. The surgeon should remember that the innominate vein crosses the trachea at the level of the suprasternal notch, and that incising this vessel will probably make tracheotomy unnecessary.

Too low a tracheotomy will cause much discomfort because the tracheotomy tube will catch on the suprasternal notch. Movements of the neck in flexion and extension during the post-operative period will tend to dislodge the tracheotomy tube (as the trachea rises and falls into the thorax during these movements). In babies, a low tracheotomy is all too easily dislodged, because babies roll their heads about much more than adults.

Too high a tracheotomy will be uncomfortable because the tracheotomy tube will irritate the cricoid, which has to be tilted up and down to tense and relax the vocal cords, and the metal tube may, indeed, cause some necrosis of that cartilage by rubbing against it.

The average *satisfactory level* for a tracheotomy opening is at about the second or third tracheal cartilage.

In an *emergency* the trachea may be incised longitudinally. A pair of forceps is inserted to divide the lips of the incision and the tracheotomy tube inserted, with the introducer. The introducer is withdrawn from the tracheotomy tube. The operation has now been completed. It only remains to tie the tapes attached to the tube behind the neck, to insert the inner tube, and to suture the incision.

If an endotracheal tube has been inserted, and there is, therefore, no imminent danger of asphyxia, it is much more satisfactory to cut a small round opening in the trachea to fit the tube that it is proposed to introduce. A small bladed knife is inserted through the fibro-elastic-muscular tracheal wall well above the selected ring. A curved incision is made to include the selected ring, but leaving a round disc, still hinged by the cartilage on one side. This lid-like disc is raised and held firmly in forceps, and the hinge cut through. This prevents loss of the disc into the bronchi should the patient take a violent inspiration at the wrong moment.

Precautions

If there has been severe unrelieved obstruction, the surgeon must be prepared for violent expiratory movement to expel mucus, or purulent bronchial exudate. This will happen almost immediately the trachea is opened. The surgeon must be prepared for this, and should keep his head out of the way, and should always wear a *cellophane mask*. It is preferable to *wear glasses;* plain glass lenses if necessary, to protect the eyes. This

is important if the obstruction is due to diphtheria or some other infection. In poliomyelitis the tracheotomy is usually performed on a patient with paralysed respiratory muscles, but, even so, it is wiser to take the precautions described. If an endotracheal tube has been inserted before the tracheotomy is performed, the patient's respiratory relief will not be as violent as it might otherwise have been. The endotracheal tube is

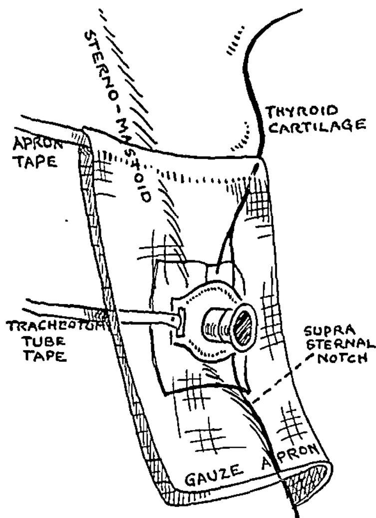


FIG. 85. Use of gauze apron to protect tracheotomy opening (For clarification, the tracheotomy tube is shown diagrammatically through the gauze apron In actual practice it would be covered by the apron.)

withdrawn as the tracheotomy tube is inserted. Suction is applied to the tracheotomy tube by means of a *rubber catheter that will pass easily down the tube, and collected* mucus removed from the trachea and main bronchi.

A piece of tulle gras (vaseline gauze) is placed under the flat plate of the tracheotomy tube next to the skin. A gauze apron is placed over the opening of the tracheotomy tube to prevent inspiration of dust (Fig. 85). *Care should be taken to prevent inhalation of* cold air during the journey back to the ward.

Types of Tracheotomy Tube (Fig. 86). The older types were made of *silver*, had a fixed plate, and the tubes were curved so as to form part of a segment of a circle. These

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If, because of obstruction below the cricoid involving the upper trachea, low tracheotomy must be performed, the same technique is adopted. The surgeon should remember that the innominate vein crosses the trachea at the level of the suprasternal notch, and that incising this vessel will probably make tracheotomy unnecessary.

Too low a tracheotomy will cause much discomfort because the tracheotomy tube will catch on the suprasternal notch. Movements of the neck in flexion and extension during the post-operative period will tend to dislodge the tracheotomy tube (as the trachea rises and falls into the thorax during these movements). In babies, a low tracheotomy is all too easily dislodged, because babies roll their heads about much more than adults.

Too high a tracheotomy will be uncomfortable because the tracheotomy tube will irritate the cricoid, which has to be tilted up and down to tense and relax the vocal cords, and the metal tube may, indeed, cause some necrosis of that cartilage by rubbing against it.

The average *satisfactory level* for a tracheotomy opening is at about the second or third tracheal cartilage.

In an *emergency* the trachea may be incised longitudinally. A pair of forceps is inserted to divide the lips of the incision and the tracheotomy tube inserted, with the introducer. The introducer is withdrawn from the tracheotomy tube. The operation has now been completed. It only remains to tie the tapes attached to the tube behind the neck, to insert the inner tube, and to suture the incision.

If an endotracheal tube has been inserted, and there is, therefore, no imminent danger of asphyxia, it is much more satisfactory to cut a small round opening in the trachea to fit the tube that it is proposed to introduce. A small bladed knife is inserted through the fibro-elastic-muscular tracheal wall well above the selected ring. A curved incision is made to include the selected ring, but leaving a round disc, still hinged by the cartilage on one side. This lid-like disc is raised and held firmly in forceps, and the hinge cut through. This prevents loss of the disc into the bronchi should the patient take a violent inspiration at the wrong moment.

Precautions

If there has been severe unrelieved obstruction, the surgeon must be prepared for violent expiratory movement to expel mucus, or purulent bronchial exudate. This will happen almost immediately the trachea is opened. The surgeon must be prepared for this, and should keep his head out of the way, and should always wear a *cellophane mask*. It is preferable to *wear glasses;* plain glass lenses if necessary, to protect the eyes. This

If the tracheotomy tube becomes displaced within 24 or 48 hours after it has been inserted, it may be very difficult to replace the tube in the ward. Force should not be used. The patient is sometimes in great distress, with froth and blood at the wound in his neck. If force is used, the metal tube may well be forced down the sides of the trachea into the cervical mediastinum causing more bleeding. The following technique is very practical and useful. A male rubber urethral catheter (about No. 6) is lubricated with glycerine or liquid paraffin. Its end is passed into the tracheotomy wound until it enters the trachea. Such a catheter cannot possibly be forced down the sides of the trachea. It is easy to determine that it is in the trachea, because it will move up and down easily if it is in the right place. The widened end of the catheter is cut off. The tracheotomy tube is fitted over the catheter and run into the trachea "on the tramline."

Cleaning the tracheotomy tube is simple. The inner tube is removed if the tube is of silver, and this is cleaned and returned. Rubber tubes have to be removed and exchanged for clean ones. *Feathers* are found in old tracheotomy sets. They were used for cleaning. It is unwise to use them whilst the tube is in the trachea. Pieces of feather can easily be inhaled, and lost. Feathers should be removed from tracheotomy sets.

Suction of mucus. A thin catheter, attached to the suction pump by the bed, is threaded down the tracheotomy tube at regular intervals. At first this may be necessary every half hour, or more frequently still. All depends upon the state of the patient, and the amount of bronchial secretion or inflammation. After a week or two, in suitable uncomplicated cases, suction can largely be dispensed with, and if the tracheotomy has to be semipermanent, or permanent, the trachea and bronchi rapidly adapt themselves. When the tracheotomy tube is connected with a breathing machine, the tracheotomy tube is disconnected at intervals for suction.

On return to full consciousness after a tracheotomy, a patient will be both relieved and apprehensive. If he has been aware of obstruction to his airway for several days, relief will temporarily overwhelm anxiety. Inability to talk, and to make known his wants, causes great distress. A pad and pencil will give considerable relief. Sometimes the relief of obstruction will not be perfect. There may be bronchitis, or other lesions in the thorax, to complicate matters. There may be extensive fixation, or inflammation in the larynx or neck. Any limitation of the full, free movements of the trachea causes discomfort, which is unrelieved by tracheotomy. Even after the restoration of a good airway by a successful tracheotomy, patients sometimes show distress until the medullary centres have compensated for the prolonged pre-operative respiratory distress. If obstruction has been present for a very long time prior to tracheotomy, and the medulla has been used to a high CO_2 content in the circulating blood, *respiration may become shallow or even cease* after tracheotomy. The respiratory centres, for the time being, do not receive sufficient stimulus, because of the low CO_2 content of the blood, after relief from long-standing chronic cyanosis. In such cases, a little CO_2, mixed with oxygen, should be administered. A careful check by the nursing staff on the respiratory depth and rate will be necessary for a few hours after operation.

Some patients adapt themselves well to a tracheotomy tube, even if this has to be retained for long periods. Others find it difficult to tolerate at first. All seem to adapt themselves very well in the course of time.

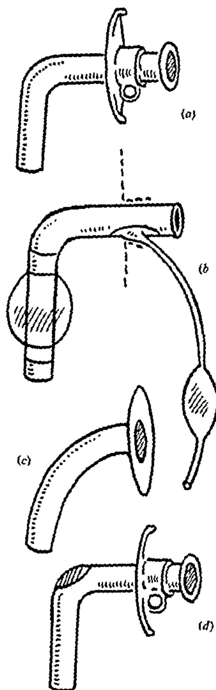


Fig 86 Tracheotomy tubes

(*a*) Silver angled tracheotomy tube, with adjustable plate.

(*b*) Rubber tracheotomy tube for use in intermittent positive pressure respiration

(*c*) Diphtheria tracheotomy tube (silver).

(*d*) Silver angled tracheotomy tube with opening above the bend for laryngeal breathing.

were fairly satisfactory for emergency tracheotomies in diphtheria, but are not as satisfactory as the *angled tracheotomy tubes* now in use. These also are made of silver, and have a moveable plate which can be fixed at any point. This is an enormous advantage, because it enables the plate to be moved if inflammation in the neck causes swelling, and so prevents displacement of the tracheotomy tube out of the trachea.

These tubes can be made with an *opening* on the upper surface of the convex side of the angle of the tracheotomy tube. This type is used if it is desired to test the patient's recovery before withdrawing the tube. Patients often dread the removal of the tube. They can be reassured if the tube already in the trachea is exchanged for a new tube, of exactly the same size, made with one of these openings. The patient can then practise talking by closing the mouth of the tracheotomy tube in the neck with the finger. This causes all the air inspired and expired to pass through the larynx. A wooden cork can then be supplied, and the patient can be encouraged to carry on for periods of time breathing normally through the larynx without the use of the tracheotomy tube. Later, as she gets used to it, she can wear the cork in at night, until everyone is satisfied that the normal airway is adequate, and the tracheotomy tube can be withdrawn and the opening allowed to close.

The third type of tracheotomy tube is the type used for intermittent positive pressure respiration in respiratory paralysis from poliomyelitis or virus polyneuritis. This is shaped like the angled tube described above, but it is *made of rubber* and has an inflatable cuff. The tube is inserted and the cuff blown up. The tube is then connected with the breathing machine. These tubes are made with a moveable rubber plate with eyelets for attaching them to tapes.

Ordinary tracheotomy tubes are, of course, sometimes made of rubber, and some patients find them more comfortable than silver tubes. The advantage of silver is that the tubes last well, stand boiling, are easy to clean, do not corrode, and do not irritate the skin.

The tracheotomy wound closes rapidly. It is quite difficult to keep it open once the tube is dispensed with. There is, as a rule, very little stenosis. The wound heals well. The scar is often unnoticeable, but can be excised later if unsightly.

(b) *People who swallow foreign bodies deliberately:*

(1) Patients in mental hospitals.

(2) Prison inmates.

(3) Exhibitionists at Fairs.

When trying to assess a patient's symptoms in the receiving room, or casualty department, this knowledge can be helpful.

Accidents. Natural teeth are themselves highly sensitive to contact with all foreign bodies. Patients with partial paralysis or paraesthesia of the mouth and pharynx, or with loss of protective taste (chorda tympani), or with very large old-fashioned dental plates, or old people without teeth, who gulp their food, will all be more liable to swallow bones than would people with normal mouths and teeth.

Infants tend to put small brightly-coloured foreign bodies into their mouths, when mistaking them for sweets, or investigating their shape and texture. Some older children seem to like holding things in their mouths.

Tailors, dressmakers, and housewives put pins in their mouths whilst both their hands are busy. Shoemakers and box makers hold nails in their mouths when working.

Accidents sometimes happen to quite ordinary people. Three recent examples will suffice.

(a) A young lady concert pianist, eagerly rising from the table to answer an expected telephone call from her fiancé hurriedly swallowed a mouthful of pigeon pie as she lifted the receiver. She stood transfixed. A sharp piece of bone, 1½ in. long, had become firmly impacted in her upper œsophagus with its long point through into the mediastinum. She had to be hurried to hospital where it was removed with difficulty.

(b) A young man was admitted to hospital having swallowed half-a-crown. The coin was impacted in his upper œsophagus, and was giving him pain. As this was an unusual coin to swallow, he was asked how it had happened. He said he was drinking a fizzy lemonade, but found it too gassy. His wife, he said, was Italian, and she advised him to put a piece of silver in his glass "to bring off the bubbles." The only silver he had in his pocket was half-a-crown. Her advice was effective, and as he was very thirsty, he was able to "polish off" his drink. Unfortunately he had forgotten the half crown.

(c) A husband attended the receiving room on Christmas Eve complaining of a pain in his chest. He asked for something to relieve it. He said it felt rather like a razor blade. He was regarded with much suspicion because his answers were very vague and evasive, and it was Christmas Eve. He was sent away; but returned with the same story. Eventually he said that he had actually swallowed a razor blade. He was not believed, but was X-rayed. He had a double-edged razor blade impacted behind his aorta. After it had been removed, early on Christmas Day, the full story came out on the flood of his relief. He had been spending the night away from home, unknown to his wife. In the morning he had sought to shave. He had been unscrewing a lady's razor, whilst holding a new blade between his lips. His thoughts were far away. The lady, feeling cheerful, had come in behind him, and slapped him on the back in a friendly way. This made him jump, and he swallowed the blade.

In all three cases the patient's attention had been diverted during the act of swallowing.

FOREIGN BODIES IMPACTED IN THE ŒSOPHAGUS

Swallowed foreign bodies are always potential coroners' cases. The danger may be immediate, reasonably urgent, or late. The following examples illustrate these points.

(*a*) A little boy of four years *swallowed a halfpenny* which his mother had given him to buy sweets. Two years later, when he was six, he vomited blood and died. His aorta had been perforated. The remarkable thing was that the impacted coin had given rise to so few symptoms, and that the boy had been able to take ordinary food, and was well nourished (Grey Turner).

(*b*) As a registrar on full duty at London Hospital, the writer was asked to see a middle-aged woman with a *large stewed mutton bone* impacted in the upper œsophagus. She was cyanotic, salivating, and in great distress, and obviously dying.

A hasty look at the larynx under extremely difficult conditions showed that the trachea *was severely compressed. A large mass of meat and bone was completely obstructing* the œsophagus and cervical trachea. A very low tracheotomy might have helped, but in her congested state might have killed her. It was impossible to pass an endotracheal tube. The removal of the mass gave relief after intermittent positive pressure respiration with oxygen.

The above are exceptional cases; but patients with foreign bodies impacted in the œsophagus should always be admitted to hospital, and the foreign body removed as soon as possible. There is usually adequate time for preparation. It is not wise to leave the patient until the morning. With every hour there may be more œdema, making identification of a very small sharp object more difficult. The object may pass further down the œsophagus, cause more laceration, and bleeding, and increase the risk. If the foreign body has been present in the œsophagus for a day or two, the damaged œsophageal walls can make the operation for removal extremely difficult. Later still, perforation, and cellulitis in the mediastinum may complicate the problem considerably. The sooner the surgeon can take the foreign body out, the easier it will be.

The management and removal of foreign bodies impacted in the œsophagus is rewarding and interesting work. The patient arrives in a distressed state and returns to normal activity in a day or so if the foreign body has been carefully removed. Failure, on the other hand, usually ends, sooner or later, in death.

Foreign bodies most commonly lodge in the lower pharynx or upper œsophagus, roughly between C.6 and T.4. Considering how often the average adult or child finds bones in a mouthful of fish or meat, and how often other small metal objects are held in the mouth, *it is really surprising how comparatively seldom such things are actually swallowed* by mistake. For this we have to thank the protective sensory receptors (tactile, pain, and taste) of the palate, gums, teeth, lips, cheek, and tongue.

Pre-disposing Factors

Certain groups of the population tend to swallow foreign bodies more often than others.

(*a*) *People who swallow foreign bodies by accident—*

(1) Elderly people with artificial dentures.

(2) Small children.

(3) People in certain occupations and trades

With the laryngeal mirror, the posterior third of the tongue, the epiglottis, the spaces between the epiglottic ligaments and the tongue (valleculæ), the piriform fossæ, the posterior and lateral pharyngeal walls, and the larynx, must all be carefully inspected. *Collected froth* at the opening of the œsophagus suggests upper œsophageal obstruction. A sharp foreign body sometimes causes severe spasm of the upper sphincter of the œsophagus (crico-pharyngeus muscles), or a large meat bone, with soft tissue attached, may block the upper œsophagus completely. One would expect the sign of collected frothy mucus in the piriform fossæ to be present in these cases, but if saliva and mucus can escape down the œsophagus past the impacted foreign body, this sign will be absent.

If the foreign body is easily accessible, it can be removed with forceps, to the immediate relief of the patient. But if the foreign body is relatively inaccessible, removal in a theatre is always wise. Anything can happen.

If the foreign body is not seen on inspection of the pharynx and larynx, and is suspected of being in the œsophagus, radiological examination should follow.

Radiological Examination

A negative X-ray of the œsophagus does *not* mean that no foreign body is present. Many foreign bodies are not opaque to X-rays. Some are only relatively opaque. Metal foreign bodies are easily seen. It is impossible to see a partially opaque fish bone in the thoracic œsophagus.

Fortunately, most fish bones impact in the cervical œsophagus where they can be seen. Here calcification in the cricoid and thyroid (and even tracheal) cartilages may cause confusion. Arthritic lipping of the bodies of the cervical vertebræ at C.6, 7 and T.1, can resemble foreign bodies. A lateral, and antero-posterior view of the cervical œsophagus should be asked for. The radiographer must be told:

(*a*) The nature of the foreign body suspected of having been swallowed.
(*b*) The probable opacity to X-rays.
(*c*) The position and level of the pain.
(*d*) The time of the accident.

The value of a Barium Swallow is doubtful. It is useful if neoplasm is suspected, but often obscures a foreign body shadow, and remnants of barium may make recognition of a foreign body down an œsophagoscope more difficult. The use of cotton-wool fibres impregnated with barium in the hope that they may catch on a non-opaque foreign body is optimistic, even though it does sometimes work. Any additional effort on the part of the patient to swallow, will automatically increase vagal peristalsis, and may cause the impacted bone to pass further down the œsophagus. Unnecessary swallowing should not be encouraged.

If the surgeon suspects that a foreign body is impacted in the upper œsophagus, a straight X-ray of the cervical œsophagus should *always* be done before œsophagoscopy is attempted. This will show the surgeon, particularly in elderly people, the presence of fixed lordosis of the cervical vertebræ at C.6, 7; T.1, due to osteo-arthritic changes. Such *fixed lordosis, spondylosis, or scoliosis*, may add greatly to the hazards of passing the œsophagoscope. Unless the surgeon is aware of these conditions, and of the rigidity of the patient's vertebræ, he may be inclined to ask for excessive muscle relaxation from his anæsthetist. What is more, he may become over-anxious and unwise, and try to force the œsophagoscope down the œsophagus. If severe fixed lordosis is present, this will

Clinical Examination

History. A good clear history is essential. One must make sure that a foreign body has, in fact, been swallowed. Fond mothers may jump to the conclusion that baby has swallowed a safety pin simply because the pin appears to be missing.

A patient who claims to have swallowed glass concealed in a bun, will help considerably if he can produce the bun with other pieces of glass still present in it. This makes his story more convincing and the pieces of glass can be tested for opacity to X-rays.

A patient who says she has swallowed part of an upper denture will assist greatly if she will produce the other fragment, because then it can be matched with the piece removed, and the operator will know that no pieces are missing.

A helpful sign is pain on swallowing. If each act of swallowing causes pain and distress, it is very likely that a bone may be present in the pharynx or upper œsophagus. The patient usually indicates one particular point below the cricoid cartilage where the pain is felt.

It is useful to ask if any meal has been eaten subsequent to the accident. People do not as a rule eat meals if each swallow produces a sharp pain, and if they think they have swallowed something dangerous, they are much too frightened.

One must be particularly cautious about this sign in very young children, even if a comparatively sharp foreign body has been swallowed.

Children have been known to finish a meal after swallowing a coin, and adults have been known to do likewise after swallowing a denture.

A pain that is continuous, and does not vary, may be caused by a scratch or tear. The bone or pin may have passed on.

Severe spasms of coughing at the time of the accident may indicate the entry of the foreign body into the larynx and lower respiratory tract.

Laryngoscopy, Pharyngoscopy

If the clinical history suggests the presence of a swallowed foreign body, it is always wise to examine the mouth, pharynx, and larynx. The site of the pain will help the examiner in his search. Fish-bones and pins have been found sticking into a tonsil, and a long thin bone may be impacted across the faucial inlet.

It must be remembered that somewhat crude efforts may have been adopted by anxious relatives or friends to dislodge a swallowed bone. Attempts to make the patient sick, to extract the bone with fingers, or to help it on with quantities of bread and potato, are quite common. Even the nasopharynx is not immune from the entrance of bones if vomiting has taken place, and visible scratches may have been caused by fingers.

If the suspected foreign body is not seen in the mouth or posterior pharynx, a laryngeal mirror should be used. A local analgesic like a 10 per cent cocaine spray, cautiously used, will often make examination easier, but as the adult patient is probably most anxious to co-operate, this is usually quite unnecessary. There is the added anxiety to the examiner that should a small foreign body become unexpectedly dislodged, the diminished sensitivity caused by the analgesic may make inhalation more likely. If, under special circumstances, an analgesic spray is used, the patient should be told not to swallow it, but to spit the bitter tasting mucus into a piece of gauze. Swallowing even small quantities of cocaine may produce pallor and faintness and add to the distress of the patient, and increase the difficulties of examination.

(*d*) Chicken, pigeon, and rabbit bones are relatively opaque to X-rays. The type that impacts is a rigid sharp splinter, about an inch and a half in length, from the shaft of a long bone.

(*e*) Mutton and beef bones are only relatively opaque to X-rays and usually have meat or gristle attached. The type most often impacted is a large piece of mixed cortical and cancellous bone from a vertebra or rib. Small, short, comparatively broad pieces pass on into the stomach.

(*f*) Pins, safety pins, wire, coins, and metal objects are opaque, and should be seen wherever they are.

(*g*) Artificial dentures are opaque if the plate is made of metal. If the plate is made of modern plastic like acrylic resin or polythene it will not be easily seen. Modern dental mechanics usually insert a small metal opaque wire in very small dentures for this very reason. The modern young woman asks for the most invisible and smallest denture possible, particularly if only one incisor is missing. These little dentures are easily swallowed.

(*h*) Buttons of some plastic materials are not opaque to X-rays. Bone, shell, or wooden buttons are relatively opaque. Metal buttons are opaque.

(*i*) The length of a bone or wire cannot be judged accurately by the X-ray shadow. It may be lying obliquely. A coin will often look much larger in the X-ray than it is—a halfpenny looks like a penny. At the London Hospital a child had two halfpennies impacted at C.7, one on top of the other, giving a shadow of only one coin.

Œsophagoscopy

The X-ray film should be on the viewing screen in the theatre so that the surgeon can refer to it. We prefer to remove all foreign bodies impacted in the œsophagus under general anæsthesia. We think, from experience, that it is safer than local analgesia. The anæsthetist inspects the larynx and passes an endotracheal tube, after induction. The larynx has already been inspected before admission to make sure that the foreign body is not across the lower pharynx or larynx. If it is, it can be removed under local analgesia in the theatre, with or without a little intravenous thiopentone. A muscle relaxant is used to make the passage of the œsophagoscope past the upper sphincter (crico-pharyngeus) easier and to inhibit peristalsis. The position of the patient is important. We prefer no fixed position. We like the patient to be lying relaxed on a soft table, without a pillow. The teeth are examined. It is nearly always easier to pass an œsophagoscope in an edentulous patient. Sharp, irregular, or loose teeth can make the operation difficult. It is as well to wear gloves to protect the hands.

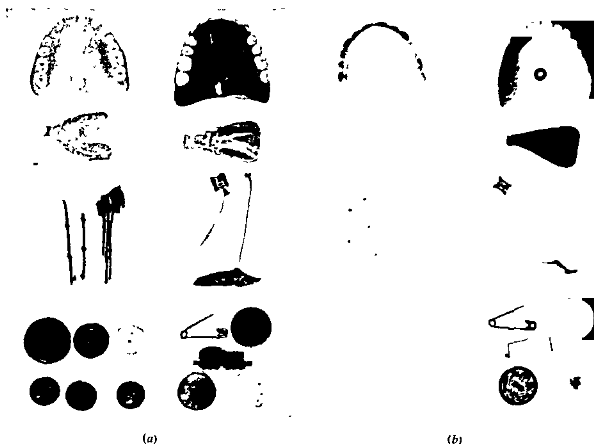
The aim of the surgeon is to pass a straight, rigid tube into the œsophagus without injuring the patient. He will require the skill of a sword-swallower. This is not as easy as it looks. If he knows his anatomy he will never do anything foolish. The œsophagus is a soft tube, with a thin muscle wall, attached above by its longitudinal fibres to the back of the cricoid cartilage. Its inner circular coat blends with the constrictor muscles of the pharynx. Except for that one attachment, the œsophagus lies suspended in the loose connective tissues of the cervical and thoracic mediastinum until it reaches the diaphragm at approximately T.10. This allows great mobility, but the relative difference between the thin soft mobile tube of the œsophagus, and the dense semi-rigid vertebræ, and the laryngeal and tracheal cartilages in front, must be appreciated.

result in tearing the œsophageal walls, and entering the mediastinum, with disastrous results to the innominate vein.

The radiographer can, therefore, supply the surgeon with two very important pieces of information:

(*a*) The position of the foreign body.

(*b*) The fixation or otherwise of the cervical vertebræ.



(*a*) (*b*)

FIG 87 (*a*) Photograph of foreign bodies commonly impacted in the œsophagus. (*b*) X-ray of same foreign bodies to show relative radio-opacity

If the X-ray reveals the presence of an impacted foreign body in the œsophagus, the patient should always be admitted for œsophagoscopy. If the X-ray does not reveal a foreign body, and it can be reasonably suspected that the foreign body is only partially opaque to X-rays, and the clinical history still suggests the presence of an impacted foreign body, then the patient should be admitted for œsophagoscopy.

Relative Opacity to X-rays of Foreign Bodies (Fig. 87)

(*a*) Some fish bones are cartilaginous and do not show in an X-ray. (Elasmobranch fishes—skate, Rock Salmon.)

(*b*) Most fish bones are only partially opaque to X-rays The type of bone that tends to impact is a semi-rigid sharp bone from the ribs, or gill cover, usually with a little soft tissue attached.

(*c*) Fish bones of any kind are unlikely to show in X-rays of the thoracic œsophagus.

œsophagoscope, forceps, and safety pin are now withdrawn as one instrument. The smooth head of the safety pin will ride quite safely outside. *If the safety pin is point* downwards it should not be pushed further or perforation of some important vessel may take place. Such safety pins are easily removed by withdrawing the pin into the œsophagoscope.

Dental Plates may cause no difficulty, if small. They should be removed with forceps made with incisor teeth because this prevents slipping. Sometimes large dentures are swallowed, and become rigidly impacted behind the aorta, or at T.5, or T.9. They may have metal hooks which catch in the œsophageal walls. It is often possible to move them into a favourable position so that they can ride safely up the œsophagus with only smooth surfaces in contact with the walls. The œsophagoscope, forceps, and denture travelling up as one instrument. The denture is usually too big to enter the œsophagoscope completely. It may be necessary to cut up the plate whilst it is in the œsophagus in order to release it. This is done with a strong saw-like forceps (Fig. 88 (*f*)), and the separate pieces are freed, and removed individually, either up the œsophagoscope, or held firmly, and the denture, forceps, and œsophagoscope withdrawn as one instrument.

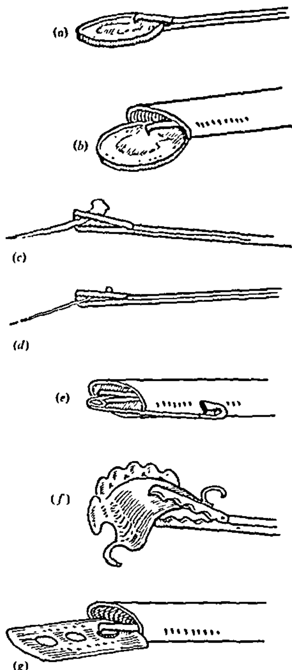


Fig 88

(*a*) *Forceps holding coin.*
(*b*) Coin in position for withdrawal with œsophagoscope.
(*c*) Forceps gripping fish bone.
(*d*) Forceps gripping pin.
(*e*) Forceps holding safety pin ready for withdrawal of œsophagoscope
(*f*) Cutting forceps breaking denture in œsophagus preparatory to withdrawal of fragments.
(*g*) Forceps holding open razor blade firmly against lip of œsophagus preparatory to rotation of forceps to fracture razor blade. Fragments are then withdrawn up œsophagoscope.

Examples

(*a*) A colleague at London Hospital removed a large hooked plate from a man aged 30. It was tightly impacted behind the heart and he had to cut it up and extract it with the greatest of difficulty, and with immense skill and patience. When the patient had recovered and asked for his denture, his first reaction was to claim damages from the surgeon for the injury to his plate.

(*b*) A woman of 52 swallowed a large dental plate when eating pork. She actually

The vertebræ are not arranged in a rigid straight line. A glance at a skeleton or X-ray will show that. *Particularly in the cervical region the curvatures are of the greatest* importance to the would-be passer of an œsophagoscope. The skull flexes and extends at the atlanto-occipital joint. Rotation movements take place between C.1 and C.2, with the dens of the axis acting as a pin pivot. The bodies of the cervical vertebræ curve *forwards convexly from C.1 to C.7, and approximately at C.7, T.1, the point of greatest* convexity is reached. The thoracic vertebræ then fall back along the posterior wall of the upper thorax and lie with the concavity of the curve forwards.

The skull must therefore be extended as far as possible at the atlanto-occipital joint, *the cervical vertebræ must be straightened out in line with the upper thoracic vertebræ.* In this way the greatest point of convexity at C.7, T.1, is reduced, and with a muscle relaxant to prevent tonic contraction of the upper œsophageal sphincter, the passing of the œsophagoscope is a fairly simple smooth manœuvre. The patient should be adjusted *to the œsophagoscope. The œsophagoscope should never be forced down the patient.* Œsophagoscopy is never really easy. One can never afford to be careless. If lipping arthritis, fixed lordosis, or spondylosis, are present, great care has to be taken, and much skill and patience are often needed. Simply bending the neck backwards will only increase the difficulty *of passing an œsophagoscope, and using* a sand bag under the shoulders will make it impossible.

This positioning for œsophagoscopy applies only to foreign bodies impacted in the upper œsophagus. If it is necessary to explore the *lower œsophagus*, the thoracic spine is raised, from underneath by an assistant, at about the level of T.4, 5, by about a handbreath, and the back of the skull lowered just a little to compensate.

For foreign bodies just inside the œsophageal opening, a laryngoscope can be used. *The cricoid is lifted with the anterior lip of the laryngoscope and the foreign body is seen* and easily removed. A short œsophagoscope is very useful. We frequently use a very short one, with a long flattish anterior lip, for use in the upper œsophagus. The operator should remember that the œsophagus passes slightly to the left of the middle line into the thorax, and he may find that *introducing the instrument down the right piriform fossa* has advantages for this reason

Coins nearly always impact at C.7, T.1. They are grasped with forceps fitted with small metal incisor teeth to prevent slipping (Fig. 88 (*a*)). The coin is held firmly, withdrawn up to the end of the œsophagoscope as far as possible and held rigidly. The œsophagoscope, forceps and coin, are now brought out, gently and slowly, as one instrument (Fig. 88 (*b*)).

Bones, Pins, and Wires are best grasped with flat-ended forceps with transverse ridges on the inner surfaces of the blades (Fig. 88 (*c*)). Bones can be turned, their buried points released, and the whole bone brought, with its sharp-cutting points, right into the safety *of the lumen of the œsophagoscope.* Whence it can be removed by withdrawing the instrument, grasping the bone, from the tube; or the œsophagoscope, forceps and bone, can be removed as one instrument.

Safety Pins give much less trouble than would first appear. They are quite commonly swallowed. They nearly always lie point upwards because they cannot travel far point downwards. The point is eased out of the œsophageal wall by holding the pin with the forceps. When the point is clear, it is brought within the œsophagoscope and held firmly. This automatically brings the head to the outside of the œsophagoscope (Fig. 88 (*e*)). The

(*b*) Before the surgeon starts, he should make sure that his forceps and suction tube are longer than his œsophagoscope. It is customary for instruments to be numbered and lettered in sets, but it is still disconcerting to be handed a sucker or forceps that will not reach.

(*c*) As soon as a small elusive foreign body is seen, it should be grasped by forceps, and removed. It is always tempting to show the foreign body to everyone in the theatre; but prolonged delay may cause the surgeon to lose sight of it. He should remember that the effects of muscle relaxants do not last long.

(*d*) Evidence of laceration or bleeding should be noted by the examiner, particularly when no foreign body is found, because these signs may indicate that an object has passed on into the stomach.

(*e*) If it is known that a small sharp opaque foreign body is present in the œsophagus, and it cannot be found, it may have penetrated the wall. Any small lesions should be carefully inspected with the sucker. If necessary, a repeat œsophagoscopy a day or two later will sometimes reveal a small patch of inflammation at the site of entry of the object, and it may then be possible to extract it.

(*f*) The possibility of swallowed foreign bodies being in the stomach or bronchi must always be considered.

(*g*) The operation should be as gentle and patient as possible, and the surgeon should aim at passing the œsophagoscope without injury to the œsophageal walls. Injury to the œsophageal walls by forcing the œsophagoscope past obstructions will only make the operation more difficult.

After Treatment

(*a*) Penicillin should be ordered systematically if perforation is known to have taken place, or lacerations are present. Pain and pyrexia should be noted. In all our experience of foreign bodies impacted in the œsophagus, only one case necessitated drainage by thoracotomy for mediastinitis with an œsophageal leak. It is indeed quite surprising how well the œsophagus stands up to injury. It is sad to have to state that the œsophagoscope is much more likely to injure the œsophageal walls than is the foreign body.

(*b*) If the walls of the œsophagus have not been injured, the patient can return to normal diet as soon as he has recovered from the anæsthetic. If there has been laceration of the walls, a restricted diet of water and other fluids may be a wise precaution for a day or two. If there is a big tear in the œsophagus, or much laceration and œdema, a Ryle's tube should be passed through the nose into the stomach, and feeding with fortified milk continued until it is estimated that healing has taken place (10–14 days).

(*c*) As a precaution, all patients should be told to report any pain on swallowing, or in the chest, after discharge from hospital.

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finished her meal, and hoped that, with reassurance, the denture would eventually pass. She was a hard-working uncomplaining woman and had little discomfort for 10 years, when she began to have difficulty with swallowing and thoracic discomfort. The plate had caused ulceration from pressure necrosis, and it was considered to be too risky to remove it during œsophagoscopy, because of the very free bleeding that took place.

Considering the close proximity of the descending aorta immediately behind and to the left, and the vena cava just in front, it is remarkable that it had not eroded through into the great vessels, as so commonly happens. It was removed successfully by thoracostomy, from the right side, *fifteen years* after it had been swallowed (Grey Turner).

A transpleural approach was made through the left seventh interspace, via an incision extending from the edge of the sternum right back to the edge of the erector spinæ. The descending aorta completely obscured the œsophagus. After division of the inferior pulmonary ligament the œsophagus was exposed and the foreign body located just below the lung root. At the site of impaction the œsophagus felt hard and simulated a malignant new growth. An incision 1½ in. long was made in the œsophagus, between guide sutures, and the denture eased out from above with a pair of angulated forceps. The wound was sutured. A drainage tube was left in temporarily. There was no excessive bleeding, and the patient made a good recovery.

Razor Blades and Similar Sharp-cutting Objects. These can cause severe laceration of the œsophagus. When located reasonably soon after swallowing, the sharp edges of the blade can be eased away from the torn walls with forceps. The blade in the case described (December, 1953) was impacted obliquely, just behind the aorta which could be seen pulsating above it. Although the walls had been lacerated there was not enough bleeding to obscure reasonable vision. The blade was gripped with flat-bladed forceps, and gently turned until its long axis was parallel with the œsophagus and œsophagoscope. It was double-edged and there were three small holes in the line down the centre. The blade was too wide to enter the œsophageal tube. It was grasped firmly at its centre line (Fig. 88 (*g*)). The œsophagoscope was eased forward over the forceps to meet the edge of the blade. This caused the blade to be firmly wedged under the anterior lip of the œsophagoscope. The forceps were rotated until the thin steel blade fractured. The separate pieces were brought up through the lumen of the œsophagoscope. There was no additional bleeding.

This method is helpful for any similar breakable foreign body, which is considered to be too dangerous to remove whole up the œsophagus. Knife blades that obviously cannot be cut or fractured in the œsophagus can usually be turned, and either brought up the œsophagoscope, or held firmly so that the cutting edge is not in contact with the œsophageal wall, and the blade, forceps, and œsophagoscope removed as one instrument.

It is remarkable how the œsophagus tolerates cutting edges. *Impaction* is the primary cause of the damage Peristaltic movements, muscular spasm, respiratory excursions, pulsation of the great vessels, all produce movement, which rubs the œsophageal wall over the cutting edge.

Comments

(*a*) If a small foreign body, thought to be impacted is not seen during the passage of the œsophagoscope, great care should be taken to look for it during the *withdrawal* of the œsophagoscope. Just as in diagnostic œsophagoscopy, the walls can be more easily inspected as they fall away from the tube during slow withdrawal.

in such cases, for the surgical evacuation of a residual collection of sterile pus and sloughed gland tissue. It may be enough to incise over the area of maximum tenderness but more often the lateral surface of the gland should be freely exposed by reflecting a cheek flap. Any collections of pus are then separately incised and drained by short incisions parallel to the branches of the facial nerve. There is little risk of a fistula resulting.

In earlier days this was often in the elderly a fatal disease with a mortality rate of 20 per cent or more, but both the incidence of this post-operative complication and its mortality have now fallen to very low levels.



Fig. 89. Parotid sialogram in a case of recurrent infection (Dr R E Steiner)

RECURRENT PAROTITIS

Clinical Features. This condition is encountered not infrequently in children as well as in adults (especially women). It is characterized by recurring bouts of swelling and tenderness of the gland on one or on both sides. There may be exacerbation of the symptoms on taking food, but the swelling always persists in some degree between meals. The disturbance is seldom an acute one and it is rare to see abscess formation. The salivary flow is usually diminished and on pressure only a small amount of muco-purulent material can be expressed from the duct: it contains pus cells and epithelial debris.

Remarkably little is known about the ætiology and the pathology of this condition.

CHAPTER IV

SECTION I

DISEASES OF SALIVARY GLANDS

M. R. EWING

ACUTE SUPPURATIVE PAROTITIS

THIS is a clinical condition which is being encountered with a rapidly declining frequency.

Pathology. The infection, which is usually due to staphylococcus pyogenes and less frequently to streptococcus viridans or B. pneumococcus, reaches the gland along the lumen of Stensen's duct. This does not happen in the presence of a free flow of saliva. The parotid with its less viscid secretion and its more dependent drainage might reasonably have been expected to be less liable to infective complications than the submandibular salivary gland, but quite the reverse holds true. Being firmly contained by its investing fascia, the inflammatory exudate accumulates under considerable tension. This leads to the development of what are usually scattered patches of suppuration with necrosis of a varying amount of gland tissue. In the neglected case a large abscess cavity will result, and this may discharge into the external auditory meatus or into the temporomandibular joint. In pre-antibiotic days downward extension into the loose tissues of the neck was a sinister complication.

Ætiology. Twenty years ago, when the urgent need of adequate hydration was less well understood than it is today, this was a not uncommon complication of major abdominal surgery or of the traditional starvation treatment of hæmatemesis. It also occurred as a complication of any long and exhausting infective illness, especially in the elderly.

Clinical Features. Pain in the parotid region is the commonest symptom, but in the acutely ill patient fever and a considerable toxæmia may be the first signs. Swelling of the gland is noted at the outset but it is important to remember that reddening of the overlying skin and fluctuation are comparatively late manifestations. Pus may be expressed from the duct. The infection is often bilateral.

MANAGEMENT

Prevention. This complication can be in large measure prevented by early and adequate hydration in the post-operative period and by rigorous oral hygiene. When swallowing is, for some reason, forbidden, frequent mouth washes should be instituted and this must be one of the few conditions when even the ascetic may condone the use of chewing gum.

Treatment. At the first hint of acute parotitis the appropriate antibiotic should be given in adequate dosage. Pending the result of a swab from the duct orifice, penicillin would be the one of choice.

Should the pain and swelling persist for 36 hours or more, formal incision is usually required. Even when the acute phase has subsided under antibiotic therapy, surgical incision may still be required: it is all too common these days to overlook the necessity,

in such cases, for the surgical evacuation of a residual collection of sterile pus and sloughed gland tissue. *It may be enough to incise over the area of maximum tenderness but more* often the lateral surface of the gland should be freely exposed by reflecting a cheek flap. Any collections of pus are then separately incised and drained by short incisions parallel to the branches of the facial nerve. There is little risk of a fistula resulting.

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Remarkably little is known about the ætiology and the pathology of this condition.

There may be demonstrable stenosis of the duct orifice, for which the irritation of a badly fitting denture is often held responsible. Some are due to an infection by streptococcus viridans and in infancy the gland enlargement is occasionally shown to be an allergic manifestation. Sialography may show a dilatation of the duct system reminiscent of bronchiectasis (Fig. 89), but there is convincing evidence that "a large part of the sialectatic picture is due to extravastation of lipiodol interstitially" (Patey and Thackray, 1955).

Treatment. Care of the teeth and dentures should be above reproach. Every effort should be made to empty the gland by carefully stripping the duct forwards with the finger after every meal. Any hint of stenosis of the duct orifice is best dealt with by intermittent dilatation using graduated lacrimal duct probes, or alternatively, by slitting up the duct opening. The symptoms may justify radiation of the entire gland in the hope of arresting completely its secretion.

SALIVARY CALCULUS

Calculus disease is encountered not infrequently. One might have expected the incidence of the disease to follow the same distribution as the occurrence of primary infection, but the submandibular salivary gland, in which primary sialadenitis is distinctly uncommon, is affected 40 times as frequently as the parotid. Presumably the more mucoid character of the secretion of the former is in some way responsible. There is usually no obvious cause for stone formation: the finding of a foreign body as a nucleus makes it a museum curiosity. The presence of a stone leads to intermittent obstruction and infection. Dilatation of the duct system and destruction of the gland tissue may result.

The calculi are almost invariably radio-opaque.

A. Submandibular Calculus

A stone may be found in the gland, in the duct, or at the sub-lingual papilla. They are often multiple and facetted and usually dirty-grey, rough and crumbling.

Clinical Features. As soon as the patient takes food the gland becomes swollen, painful and tender, but subsides rapidly thereafter: often, however, it remains appreciably enlarged even between meals. The calculus can usually be seen or felt (best bi-manually) either at the papilla or in the accessible portion of the duct, but when situated in the gland or in the proximal part of the duct it may escape detection. There is always the risk of an acute flare-up with œdema of the loose tissues in the floor of the mouth and below the mandible, or of a purulent adenitis and abscess formation. Under these circumstances the calculus can seldom be felt Occasionally a large duct calculus may ulcerate through into the mouth. It may be possible to express slightly turbid or even frankly purulent material from the duct; alternatively, every attempt to produce a flow of saliva fails.

It is often difficult to identify and to probe the duct. A sialogram may show a calculus and usually too a dilatation of the duct system An X-ray with an intra-oral dental film is a distinctly easier diagnostic procedure

Treatment

(*a*) *A Calculus in the Accessible Part of the Duct* This can readily be removed under local anæsthesia in an out-patient clinic. When the calculus has been identified with precision and fixed under one's finger, the minimum of local anæsthetic (with adrenaline

added to lessen hæmorrhage) is injected into the overlying mucosa. A bold cut is now made down to the calculus which is extracted without difficulty.

(*b*) *Calculus in the Posterior End of the Duct.* Access to this portion of the duct in a sensitive patient is often difficult. The stone tends to be elusive and its position may be obscured following the infiltration of the local anæsthetic. General anæsthesia through an endotracheal tube and with the fauces packed is a wiser procedure. The duct should then be fixed by under-running it with a silk stitch behind the stone, which is then reached by a bold incision through the mucosa in the floor of the mouth.

(*c*) *Calculus in the Gland.* The only satisfactory way of dealing with a calculus at this level is by excision of the gland through an incision below the mandible. The incision should be adequate in length for bleeding from the facial vessels may be troublesome and the nearness of the fifth and twelfth nerves makes careful dissection obligatory.

B. Parotid Calculus

This is encountered very infrequently. If it is situated far forwards in the duct it can easily and safely be reached from the mouth. If in the masseteric portion, it must be exposed and removed through an incision in the cheek, the duct and its coverings being then reconstituted with the very greatest precision.

SALIVARY FISTULA

A. Internal. An internal fistula is established whenever a stone in the accessible part of Wharton's or of Stensen's duct is removed from inside the mouth. It causes no trouble subsequently.

Although in the resection of a carcinoma of the floor of the mouth, the submandibular salivary duct may be in part removed, it is not necessary to make any formal attempt at mucosal implantation of the divided proximal end. It is true that chronic enlargement of the gland (and in the case of a mid-line lesion this may be bilateral) may be noticed, and may persist after operation, although usually without symptoms. Similarly, it is unnecessary to worry about the fate of the parotid duct when resecting a cancer on the buccal mucosa.

B. External. Although a good deal has been written about the management of salivary fistulæ, they are in practice distinctly uncommon.

A salivary collection in the wound or a small leak in the early post-operative period is a not infrequent happening after the excision of a parotid tumour, or following excision of the lower pole of the gland during a radical neck dissection. This always dries up quickly and spontaneously.

External injury to the main duct of the parotid, whether due to external injury or following the removal of a stone, may be followed by the development of a most intractable salivary leak on the cheek. This is especially so when the fistula is in the masseteric part of its length. A variety of surgical manœuvres has been described to divert the fistula back into the mouth, but even in the absence of any obstruction at the buccal end of the duct, disappointing relapse is a frequent happening. Often the easiest way of arresting the salivary secretion is by the careful administration of an adequate dose of irradiation to the gland. Interruption of the para-sympathetic secretory fibres by division of the auriculo-temporal nerve is not only technically difficult but too uncertain in its effect to be of real value.

OTHER CAUSES OF SALIVARY GLAND ENLARGEMENT

EPIDEMIC PAROTITIS OR MUMPS

This is a well-recognized acute infectious illness characterized by non-suppurative parotitis and mild constitutional symptoms. The swelling usually affects each gland, although the onset is not necessarily simultaneous on the two sides. Spontaneous and complete resolution is the rule.

It is important to remember (1) that although the disease most often affects children, it occurs not infrequently in adults and (2) that although a second attack is possible, it can be accepted as a general working rule that a first attack usually affords a life-long immunity.

Swelling of one or other of the salivary glands is a manifestation of a variety of clinical conditions, many of which are of such ætiology that their eponymous titles seem likely at the moment to endure. It is, however, highly probable that further experience will prove many of them to be but variants of a basic disorder. It has certainly been shown recently that several of them seem, at least histologically, to follow the same essential pattern.

MIKULICZ' DISEASE

There occurs rarely a benign, self-limiting disease in which the only symptom is enlargement of the salivary and of the lacrimal glands. Histological examination reveals atrophy of the gland acini which become separated by lymphoid tissue. There is no sign of disease elsewhere in the body. The causation is unknown.

MIKULICZ' SYNDROME

In a variety of general affections of the lympho-reticular system, swelling of the lacrimal and of the salivary glands is a prominent clinical feature: it may occasionally be the first (Fig. 90). The primary disease may be Hodgkin's disease, lymphosarcoma or lymphatic leukæmia. The swelling in such cases will usually respond to a course of external radiation, but the patient almost always succumbs eventually from the primary disease of which the enlargement of the salivary and lacrimal glands is no more than an incidental clinical manifestation.

SARCOIDOSIS

Enlargement of the parotid or of other salivary glands may be one of many manifestations of sarcoidosis. X-ray examination of the chest or aspiration biopsy of the parotid or liver may help to establish the diagnosis. A good many cases labelled Mikulicz' disease are probably of this nature.

UVEO-PAROTITIS

It is exceedingly doubtful whether or not this exists as a distinct and separate clinical entity: careful documentation will usually prove them to be cases of sarcoidosis. These patients come first to the ophthalmologist suffering from a non-specific inflammation of the pigment coats of the eye. Then or later they are found to have, in addition, parotid gland enlargement, but with no tendency to pus formation.

S. SJÖGREN'S SYNDROME

In 1933 Sjögren described an interesting syndrome which now bears his name. The condition is distinctly rare, but is worth bearing in mind as a cause of salivary gland enlargement. The patient, who is almost invariably a female at or about the menopause, usually first finds her way to the ophthalmologist complaining of smarting of the eyes,



FIG 90. Mikulicz syndrome in a woman of 44. Bilateral parotid swelling biopsy showed lymphoid tissue in which were scattered residues of salivary gland tissue.

often of long duration and often too of photophobia and loss of visual acuity. She is found to have keratoconjunctivitis with a reduction in the lacrimal secretion. Later, there appears a recurrent swelling of one or more of the salivary glands (most commonly the parotid), dryness of the mouth (xerostomia), rapid dental caries, polyarthritis and absence of sweating. Occasionally the parotid swelling is the first manifestation of the disease. The swelling is commonly bilateral, the glands being firm and tender. Regression occurs without going on to pus formation. Biopsy of the enlarged gland may help in establishing the diagnosis. Histological examination reveals atrophy, round cell inflammation and fibrosis.

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wall of the oropharynx in the tonsillar region. Bimanual examination inside and outside usually demonstrates the continuity between such a tumour and the remainder of the parotid gland.

Sialography has little to offer in establishing the diagnosis. At best it shows only an area in the parotid where the normal duct system is not outlined.



FIG. 91 A typical mixed tumour of the parotid in a woman of 74 Duration—3 years

PATHOLOGY

Although the surgeon can at operation commonly demonstrate to his own satisfaction the existence of a capsule and may even speak confidently of "enucleating" such a tumour, the histologist when he comes to examine the entire gland seldom reports the finding of a convincing investing layer (the same is often true in the case of a fibroadenoma of the breast). Even where a capsule can be demonstrated it is often incomplete and it is a commonplace to see small, independent, satellite growths adjacent to, but apparently outside the main tumour mass.

The most striking feature of the tumour microscopically is the differing structure as seen in even closely adjacent fields (Fig. 92). It was this feature which, no doubt, earned

Treatment is of a symptomatic nature. A.C.T.H. can induce a temporary reduction in the size of the swelling.

SALIVARY GLAND TUMOURS

Mixed Tumour

The great majority of salivary gland neoplasms are of this type. They may occur in any of the salivary glands, major or minor. They are, however, in this country at least, encountered very much more frequently in the parotid than elsewhere. ". . . for every 100 parotid tumours, there are likely to be seen about 10 sub-maxillary tumours, 10 tumours of the minor salivary glands (of which about half will be palatal) and only one sub-lingual tumour" (Willis).

The incidence in the parotid gland seems to be greater than could reasonably be accounted for by its greater bulk in relation to other salivary glands. It is possible that its proneness to tumour formation may be in some way related to its high proportion of serous (in contrast to mucus-secreting) cells.

Mixed salivary gland tumours are met with at all ages: they are by no means uncommon in childhood or in adolescence. The peak incidence, however, is at a slightly higher age period, the patients commonly first seeking treatment in the thirties or forties.

There is no uniformly reported preponderance in either sex.

A. Parotid

Clinical Features

Commonly the patient's only complaint is of the gradual appearance of a swelling in the region of the angle of the jaw. Although the rate of enlargement is widely variable in the untreated case, it is typically very slow indeed. It is, therefore, not surprising that the tumour is characteristically painless. It is for this reason too that the skin, although it may be tightly stretched over a massive tumour, seldom ulcerates, and the facial nerve, although grossly distorted, escapes with its function quite intact. Little wonder too that the patient often comes for treatment relatively late and then only when the tumour is by its bulk causing considerable disfigurement. The average duration of symptoms before treatment in Ahlbom's series of 251 cases of salivary gland tumours at all sites, was close on 7 years. Even with the bulkiest tumours, trismus is uncommon.

The tumour is usually situated in the most superficial part of the gland and commonly towards its lower pole. It grows outwards and presents as a swelling below and in front of the ear (Fig 91). Often it is close up against the lobule or even at a slightly higher level in front of the tragus. Alternatively it may lie precisely over the angle of the jaw, or rather more posteriorly when it presents primarily as a neck swelling. Superficial tumours often seem, by virtue of their mobility, similarly to be quite distinct from the parotid and from their position suggest rather the diagnosis of a subcutaneous or even of a dermal neoplasm. They remain, however, in the untreated case, always unattached to the overlying skin

Typically the tumour is elastic and firm, but not hard. Although there may often be histological evidence of cyst formation, detectable fluctuation is an infrequent finding. The surface is usually smooth, but may be slightly lobulated.

Very occasionally a tumour in the deeper part of the gland may extend towards the fauces and cause dysphagia. It then presents as a smooth tumour distorting the lateral

are not far removed from frank carcinomata. What has probably attracted most attention of all is the finding of areas which look, at first sight, like cartilage. It was their existence, coupled with the fact that the tumours seem often to be quite separate from the gland proper, which led to the belief that they were in part mesodermal and probably of branchial origin. It is now established that this cartilage-like material is no more than a mucinous derivative of the epithelial cells of which the tumour is primarily composed.

The salivary gland tissue of the remainder of the parotid seldom shows any deviation from normal.

Diagnosis

A painless swelling of long duration and slow growth, anywhere in the parotid region, should at any age suggest the diagnosis of a mixed parotid tumour, even when the feeling of the lump suggests that it is quite distinct from the parotid gland. The error in the clinical diagnosis of mixed parotid tumour is in the region of 50 per cent (Patey, 1952). Biopsy as a method of establishing the diagnosis is subject to some limitations and has not been widely accepted as a certain way of arriving at a firm diagnosis.

Differential Diagnosis

(1) Other causes of chronic parotid gland enlargement such as recurrent pyogenic infection, calculus, sarcoidosis, Mikulicz' syndrome, parotid cyst (which may mask an underlying cancer), lipoma, angioma.

(2) Adenolymphoma—see below.

(3) Carcinoma of parotid—see below.

(4) Adamantinoma. This is another cause of a slow-growing and painless swelling in the region of the angle of the jaw. Examination of the molar region and an X-ray should immediately suggest the correct diagnosis.

(5) A lymph node enlargement—parotid, submandibular or upper deep cervical—whether due to pyogenic infection, tuberculosis, reticulosis or cancer.

(6) Branchial cyst.

(7) A superficial tumour is often considered to be a sebaceous cyst, or a subcutaneous lipoma. Such a mis-diagnosis often leads to its (usually inadequate) removal in a hospital outpatient department.

Natural History of the Tumour Untreated

If left quite alone the tumour is likely, in a matter of years (varying from one or two to very many), to become increasingly unsightly. In general, it will seldom kill the patient and is very rarely found spontaneously to assume the aggressive characters of a malignant growth.

Behaviour of the Tumour Following Surgical Removal

Experience has shown local recurrence of the tumour to be a very frequent happening following attempts at surgical excision. The percentage of failure may be as high as 20–25 per cent in the first 3 years (Ahlbom) and there is still a continuing risk of later recurrence for as long as 20 years thereafter.

The high incidence of recurrence can be attributed to two main factors:

(1) The majority of failures are due to inadequate surgical excision. We have seen above that there are commonly outlying portions of these tumours beyond the poorly

for them the epithet "mixed." It is, however, a descriptive term which is misleading in so far as it suggests that they have some of the features of a teratoma, for, it seems now to be generally agreed by pathologists that these tumours are derived solely from salivary gland epithelium and that the varied histological pattern is no more than a peculiar accident, which results from the growth in a certain environment of a modified epithelial



FIG 92 Mixed tumour of the parotid—(×95)

cell of the skin. (Similar appearances are seen, for example, in some skin, breast and sweat gland tumours.) Willis has tried to popularize the alternative title of "pleomorphic salivary gland tumours" but the term "mixed tumour" is, by now, so firmly established as to make the chance of any change at this stage most unlikely.

The cells may be arranged in all sorts of ways—in acini, in columns, in clumps, with a cribriform structure or with cyst formation. Often there are areas where the cells have epidermoid features and sometimes groups of large eosinophilic cells of distinctive appearance, mysteriously called "onkocytes." Some of the tumours, which are highly cellular and which are composed of irregular, actively dividing, poorly differentiated cells

The most satisfactory is to identify the main trunk of the nerve behind the ascending ramus and to follow it forwards into the gland. If the tympanic plate is identified the nerve will be seen 0·5 cm. below its free margin: alternatively, it can be seen as it emerges from the stylo-mastoid foramen after detaching some of the anterior tendinous fibres of the sterno-mastoid and nibbling away the lower and anterior part of the mastoid process.

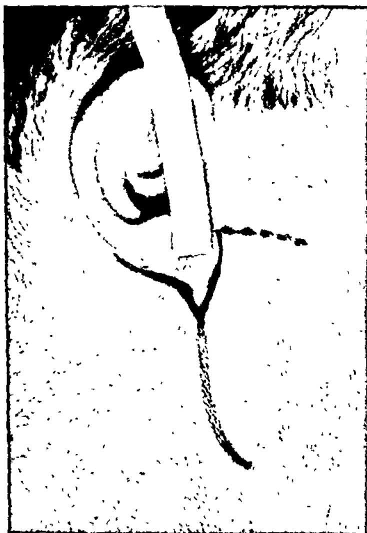


FIG 93 Good access to the dissection of the facial nerve by a Y-shaped incision If the tumour lies far forward in the gland the anterior limb can be extended forwards across the cheek.

The alternative method of identifying the cervical branch of the nerve after it leaves the gland and following it proximally is a much less satisfying manœuvre. The main trunk having been identified, the branches of the nerve are then followed forwards into the gland and as much of the latter removed as seems necessary. One may not infrequently be obliged to sacrifice a few fine filaments, but their loss leaves no disability. Pains should, however, be taken to preserve the nerve supply to the orbicularis oculi.

Some degree of facial weakness is not uncommon after operation, but this usually clears completely.

A salivary collection under the skin flaps may occur but a fistula almost never.

defined capsule. Any excision which does not clearly encompass the tumour and a protective kernel of normal adjacent salivary gland invites recurrence. Simple enucleation, with its attendant risk of spillage of tumour content, is certain to be inadequate. The restricted nature of the resection reflects in most cases the not unnatural timidity on the part of the surgeon to carry out a wider removal for fear of damage to the facial nerve.

(2) Recurrence may be single or multiple, superficial in the operation scar, or deeply placed in the gland itself. Often it is distinctly firmer and less regular than was the original tumour. Often, too, the rate of growth appears to be accelerated by operation and what was at the outset an indolent tumour, in its recurrence grows much more rapidly. The histology of the tumour usually remains, however, quite unchanged.

Treatment

(1) Conservative. Knowing that the tumour is seldom lethal and usually painless, there can be a strong argument for leaving severely alone any mixed parotid tumour in the elderly, always provided a clinical diagnosis of benignity can be made with assurance.

(2) Surgical Excision. This is undoubtedly the method of choice always provided it is adequate.

The following methods have been advocated:

(a) *Enucleation of the tumour from inside the capsule*, which is then separately seized and withdrawn. This can only result in an incomplete resection and has nothing to recommend it.

(b) *Removal of the tumour along with its capsule and an adequate layer of surrounding parotid gland* This would seem to be the method of choice. It is becoming increasingly apparent that if this resection is to be as wide as the high risk of local recurrence requires, the precise identification of the seventh nerve in the gland and its subsequent careful avoidance is an essential preliminary. The natural extension of such an operation, to give the complete assurance of no further recurrence, would be a formal removal of the entire gland, always provided experience showed that such a resection could be carried without a facial paralysis. Recent writings certainly suggest that parotidectomy is becoming increasingly widely practised (Redon, 1945). Whether or not the average parotidectomy is ever a truly total one is debatable, but it is true at least that tumours almost always occur in the more accessible portion of the gland and that this segment can readily be removed as a complete structure.

The argument in favour of a parotidectomy is even stronger, in the treatment of a recurrent tumour. Excision should then properly include the scar of the earlier operation.

Operative Technique. The first requirement is an adequate exposure. A Y-shaped incision, the diverging limbs of which embrace the attachments of the lobule, allows the auricle to be turned upwards and the dissection to be continued as far as the cartilage of the external meatus. The lower limit of the incision comes forwards below the angle of the mandible (Fig. 93) and if access is required to the anterior portion of the gland the upper anterior limit of the incision can be carried horizontally forwards across the cheek.

The second requirement is a relatively bloodless field, a matter of considerable difficulty in a vascular structure such as the parotid. However, by diligent suction and by posture, and on occasions, by use of hypotensive drugs, a relatively dry operation field can be achieved.

For the identification of the facial nerve a variety of methods has been suggested.

(3) *Radiation alone.* Some radiotherapists believe operation to be entirely unnecessary, but this view is certainly not shared by surgeons.

PROGNOSIS. In Radiumhemmet (1950) only 1·7 per cent of recurrences were noted after 5 years in a group of 540 benign and semi-malignant salivary gland tumours treated by limited surgical excision combined with radiotherapy.

B. Mixed Salivary Gland Tumours at Sites other than the Parotid

In general, tumours in these situations have the same clinical and histological features as the more commonly occurring tumours in the parotid. It is, however, true that a significantly higher proportion of them behave like malignant tumours, as shown by aggressive local extension or by spread to the regional lymph nodes. The main symptom is of a painless swelling which has increased slowly in size over a matter of years. Each presents, however, peculiar problems in diagnosis and in treatment which merit mention separately.

(1) SUB-MANDIBULAR

Differential Diagnosis. The commonest cause of sub-mandibular salivary gland enlargement is obstruction associated with calculus. The case with recurring painful post-cibal enlargement is unmistakable, but, especially when the calculus is in the gland itself, there may be a chronic persisting swelling.

The sub-mandibular salivary gland bears such a very close anatomical relationship to the adjacent lymph nodes, that it is often exceedingly difficult to decide to which of the two a swelling is due. An essential part of the clinical examination is a careful review of the mouth, the antrum, and the skin of the face, lips and neck, to exclude the presence of a cancer at any of these sites, bearing in mind a minute and insignificant melanoma as an easily overlooked primary.

The sub-mandibular region should be remembered as an uncommon site of tuberculous lymph node enlargement (Fig 94).

In the absence of any other cause of lymph node or of salivary gland enlargement, the diagnosis narrows down to a choice between mixed tumour and one of the reticuloses. A long history favours the former, and any hint of node enlargement elsewhere, the latter.

Biopsy will decide the issue but if the operative findings point to the diagnosis of mixed tumour, a formal excision forthwith (see below) will save a second operation.

Treatment. Simple enucleation of the tumour is followed by a disturbingly high percentage of local recurrence: the tumour can be removed adequately only by sacrifice of the entire gland.

(2) PALATE

A mixed tumour of one of the minor salivary glands of the palate usually presents as a firm (occasionally it may be slightly fluctuant), rounded, sessile tumour which has slowly increased in size over a matter of years It is usually slightly to one side of the midline and may be situated on the hard or on the soft palate. It grows beneath the mucous membrane which may, however, later become ulcerated. The wearing of a denture becomes impossible and the large neglected tumours may fill half the mouth.

Hyperæmia of the skin corresponding to the distribution of the auriculo-temporal nerve with profuse sweating in the same area, may follow parotidectomy. Usually they occur in response to some gustatory stimulus. Known to surgeons as Frey's syndrome, it tends with the passage of time to become less disabling although it seldom clears up completely.

(3) RADIATION. By and large, mixed salivary tumours are not radiosensitive. Radiation has, however, been advocated either as a pre-operative or as a post-operative measure or as the only method of treatment.



FIG 94. Tuberculous lymphadenitis involving the submandibular group of glands

(1) *Pre-operative.* It is claimed that its use before operation induces some toughening of the capsule which diminishes the risk of rupture and dissemination of its contents during removal.

(2) *Post-operative.* Following the lead of Ahlbom it has widely become the practice to follow excision by a course of post-operative radiation. This certainly led in the practice of Radiumhemmet in Stockholm to a significant lowering of the recurrence rate, and this experience has been confirmed elsewhere. Radiation has variously been given by the external method or by a buried source of radium.

mixed tumour. A superficial mobile cystic tumour in the parotid in an elderly male, of slow painless growth, is not unlikely to be an adenolymphoma but it might just as readily be a mixed tumour. Aspiration biopsy has been of value in establishing the diagnosis before operation. It may be mistaken for a caseating tuberculous lymph node.



FIG. 95 Adenolymphoma of the parotid (×80)

TREATMENT

Removal by simple enucleation is all that is required. The tough and obvious capsule makes this easy and the facial nerve is never in danger. There is no risk of recurrence or of malignancy.

Carcinoma of Parotid

Although it is convenient for the clinician to consider some of his patients separately and to give them this diagnosis, pathologists are becoming increasingly insistent on the continuity of the entire group of salivary gland tumours from the benign, through the semi-malignant (Ahlbom) to the obviously malignant.

These patients present in one of two ways:

The most striking clinical feature is the complete absence of pain.

Differential Diagnosis. The long history and its rounded appearance differentiate it from a primary squamous carcinoma of the palatal mucosa or of the more common antral cancer erupting through into the mouth. The recognition of the torus palatinus should cause no difficulty: this is a symptomless bony exostosis at the top of the arch of the hard palate, present from birth, although often first detected in adult life.

Treatment. Although these tumours can be enucleated with little difficulty, the risk of recurrence is no less than in mixed salivary tumours elsewhere. An adequate sheath of apparently healthy tissue must be sacrificed on all sides; the raw area will heal by granulation.

(3) Other Minor Salivary Glands

Mixed tumours are also encountered, but much less frequently, in the sub-lingual gland as well as in minor salivary glands, including those of the lips, buccal mucosa and tongue.

Adenolymphoma

This is much less frequent than the mixed tumour, forming rather less than one in ten of all salivary gland neoplasms. It is also known variously as papillary cyst-adenoma lymphatosum, Warthin's tumour or onkocytoma.

This variety occurs most frequently in the parotid but is also met with occasionally in the submandibular gland. Men are affected much more frequently than women, in the proportion of 6 or 7 to 1. The age of maximum incidence is 10 years or more later than in the case of the mixed tumour. Like the mixed tumour it presents clinically as a very slow growing rounded or slightly flattened, painless swelling, usually situated in relation to the lower pole of the parotid. It is seldom large, feels quite superficial and movable and seems often to be quite distinct from the parotid gland proper. There is no fixation to the skin. It may be obviously cystic and is often lobulated. Not uncommonly a similar tumour is found in the opposite gland and multiple unilateral tumours have also been reported.

Pathology

The tumour is often found to lie close to, but not obviously in direct continuity, with the parotid gland. It possesses in contradistinction to the mixed tumour, a clearly defined tough fibrous capsule and on section can usually be seen to contain many small cystic spaces, some of which may be lined with shaggy papillary fringes and occasionally milky material not so very unlike caseating tuberculous debris. The cells lining the cysts are pseudostratified columnar in type, and the stroma is packed with lymphocytes (Fig. 95). The histological picture is quite distinctive: it can readily be differentiated from a mixed tumour and there is no hint of malignancy.

It is suggested that these tumours take origin from inclusions of parotid (or of sub-mandibular) gland tubules, which can often be demonstrated microscopically in lymph nodes close to the normal parotid gland. The alternative theory that they arise from epithelial cells with distinctive characters called onkocytes which are found constantly in salivary glands after the age of 60 or so, does not now win wide acceptance.

Differential Diagnosis

It is almost impossible to differentiate clinically between an adenolymphoma and a

nature overlooked. The dense infiltration of actinomycosis has occasionally caused trouble in the diagnosis of parotid cancer.

Treatment

SURGERY

In the absence of hopelessly wide local extension or of metastases other than in the regional lymph nodes, total parotidectomy is the treatment of choice.

This usually implies sacrifice of the implicated facial nerve and to gain access to the deeper part of the gland, it may prove necessary to resect the ascending ramus, partially or completely. Any involved overlying skin must be excised and it may be necessary to make good the loss by a free skin graft. Even in the absence of palpable regional node enlargement, total parotidectomy can conveniently be combined with a standard block dissection of the neck nodes, the contents of the neck being turned upwards and the external carotid artery ligatured before the excision of the parotid is begun.

Care must be taken in any operation which involves paralysis of the upper components of the facial nerve, to safeguard the cornea from injury from the very outset. This can most readily be achieved by fusing the lid margins at both the inner and the outer canthus, leaving only a small gap opposite the pupil. The sooner this tarsorrhaphy is done the better.

RADIOTHERAPY

This should be reserved for the case which is inoperable in virtue either of wide local extension or of distant metastases. Radiation in such cases is likely to be of little more than palliative value.

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SECTION II

DISEASES OF THE LIP, TONGUE AND JAW

BENIGN TUMOURS

Mucous Cyst

Small, soft, cystic swellings are met with not uncommonly on the gingival aspect of the lip, especially the lower one. They seldom measure more than 5 mm. across but as they are constantly felt by the patient's tongue they appear to be very much bigger. They contain clear mucus and are probably retention cysts of the mucus-secreting glands in the

(1) The patient with a slowly growing, painless parotid swelling with every clinical sign of benignity who (*a*) very rarely spontaneously or (*b*) more frequently after operation develops regional lymph node or distant metastases or signs of local destructive infiltration of which seventh nerve paralysis is the most striking. It is often assumed, in such cases, that operation has in some way transformed the tumour and converted a benign into a malignant neoplasm. It seems more reasonable, however, to suggest that the tumour was a slowly growing cancer from the start. It cannot be denied, however, that surgical excision seems often to speed up the tempo of disease. It must also be freely admitted that the difference between a tumour which is clinically an actively growing and obvious cancer from the start and the mixed tumour which recurs and later metastasizes, is only one of degree: separation of one from the other is distinctly artificial.

(2) The patient who comes with what is from the very outset a manifestly malignant tumour which, if unchecked, will inevitably lead to his death.

The history in such cases is short, being measured in months rather than in years. Pain is not an infrequent symptom and dysphagia and trismus may also be present. The tumour is typically hard, and has an irregular surface, but a rapidly developing parotid cyst may in an adult conceal an underlying cancer. The skin in the more advanced stages is fixed, reddened or even ulcerated. The tumour is often fixed deeply and an X-ray may show destruction of bone in the region of the mastoid or of the external meatus. In such cases there is commonly paresis of the last cranial nerves. Quite the most striking clinical manifestation is a seventh nerve paralysis although this is not always complete: weakness of the facial muscles in a patient with a parotid tumour who has never previously been submitted to surgery is strong presumptive evidence of its being cancerous.

Enlargement of the regional nodes in the upper jugular chain (20–30 per cent of cases) points to metastatic spread and the investigation of such a case would be incomplete without an X-ray of the chest and of representative bones.

Pathology. These tumours are usually found to be richly cellular and anaplastic. Willis, however, is of the opinion that careful histological examination will always reveal in some part of the tumour the distinctive histological features of the mixed salivary tumours.

Biopsy. This will usually confirm the diagnosis in tumours of the second group, but in the first it is of very limited value, since a tumour of long standing which has then gone on to metastasise may retain its original (benign) histological picture quite unaltered.

Differential Diagnosis. In differentiating the benign from the malignant parotid gland tumour, shortness of symptoms, facial paralysis, hardness and fixity of the tumour (and, of course, evidence of distant metastases) strongly favour the latter diagnosis. A long history and an intact facial nerve are, however, by no means incompatible with a diagnosis of carcinoma.

Enlargement of the lymph nodes, which lie in and in close proximity to, the parotid gland, due to metastases from a carcinoma in the mouth or pharynx, is a cause of a hard parotid swelling which must always be borne in mind. An essential part of the clinical examination of any apparent primary parotid cancer is a careful scrutiny of the mucous lining of the mouth and throat, and especially of the nasopharynx.

Occasionally, too, the parotid may become involved by direct extension of a buccal cancer. The occurrence of a parotid swelling in such a case following successful eradication of the tumour in the mouth may be attributed to suppurative parotitis and its true

somewhat firm enlargement of the lip is apparent early in life (Fig. 96) Like lymphangiomatous tissue elsewhere it shows proneness to recurrent inflammation, which leads to a steady increase in the size and solidity of the swelling The only satisfactory method of treatment is surgical excision of all the affected tissues, followed by plastic reconstruction.

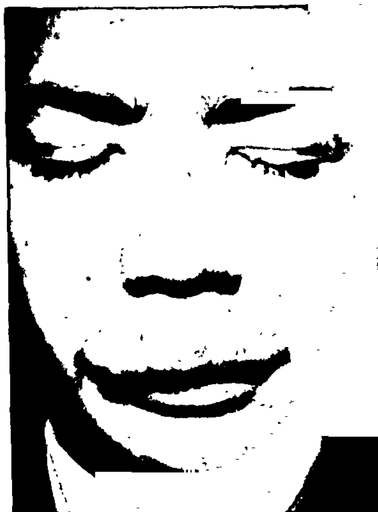


FIG. 96 Macrocheilia.

CANCER

Definition

It is customary to consider under this heading only those tumours which affect primarily the exposed parts of the mucous membrane of the upper and lower lip. In its malignancy, cancer at this site is intermediate between squamous carcinoma or rodent ulcer of the skin of the lip (both of which are relatively benign) and the malignant growths which are met with infrequently on the mucous membrane lining the inner (gingival) aspect of the lip, and occur on the buccal (cheek) mucosa.

Site

The lower lip is involved in the great majority of cases: of 293 patients, in eight only was the cancer on the upper lip (Charteris, 1946). The central and lateral portions of the

substance of the lip. As seen through the mucous membrane they have a distinctly bluish tinge: occasionally as the result of injury the mucous membrane may ulcerate.

Excision is easily carried out under local anæsthesia and recurrence is unusual, even when the cyst is ruptured during its removal. The wound is closed by one or two catgut sutures.

Papilloma

Clinical Features. This presents, not infrequently, as a greyish-white sessile warty tumour on the exposed part of the lip. Although the tumour itself is firm there is no hint of infiltration at its base. Histologically there is hypertrophy of the squamous epithelium and elongation of the rete pegs. It may be surmounted by an accumulation of keratin and, if this is a striking feature, it will clinically resemble closely the cutaneous horn seen on the skin of the face and elsewhere. In such cases there is a significant risk of malignant degeneration.

Treatment. The tumour should be removed along with a clear 2 mm. of intact mucous membrane and the base should be carefully examined histologically.

Hæmangioma

Hæmangiomata of the lip are not uncommon in infants and in young children. They may involve the whole thickness of the lip with resulting disfigurement and feeding difficulties.

Ulceration and recurrent bleeding may be a feature of the larger lesions. Spontaneous retrogression can confidently be awaited in the smaller ones, while bulky and extensive tumours are best handed over to a plastic surgeon for excision and reconstruction. Radiotherapy in very low dosage may lead to a slow obliteration of the tumour by thrombosis but if it is not given with the greatest of care there is a considerable risk of arrest of the growth of the teeth or of damage to the skin of the lip itself.

Pyogenic Granuloma

This lesion resembles histologically as well as clinically the similar lesions which are met on the tongue or on the skin. It usually follows an injury, as for example, when the lip is punctured by being driven against the ragged edge of a tooth. At the site of injury there develops a small 4–5 mm. red rounded elevation, which is tender and which bleeds on the slightest provocation. Simple excision of the tumour under local anæsthesia gives immediate relief and lasting cure.

Mixed Salivary Gland Tumour

The lip is an infrequent site of pleomorphic salivary gland neoplasms. It presents clinically as a painless tumour on the mucosal aspect of the upper or lower lip. It grows very slowly over a matter of years. It will ultimately result in considerable distortion of the lip, but even when it is very large there is surprisingly little disturbance of function. The tumour seems most often to run a benign course and experience suggests that a simple enucleation will, in most cases, result in cure.

Macrocheilia

The lip is one of the more common sites of a capillary lymphangioma. A diffuse and

destruction of the lip results. The tumour then spreads to the gingiva, to the body of the mandible and to the floor of the mouth. "Kissing" cancers, on the opposite lip, presumably by the direct implantation of cancer cells, is a very rare clinical curiosity.

Spread to the regional nodes occurs relatively late. The submandibular group is mainly affected, those in the submental area being much less frequently involved. Primary spread to nodes in the jugular chain is distinctly unusual. From a tumour on the upper lip emboli may first pass to the pre-auricular and parotid lymph nodes. A centrally situated lesion or a massive unilateral one, may spread to nodes on each side of the neck.

Distant blood-borne metastases are unusual, even in the patients who come to autopsy.

Clinical Features

The patient's first complaint is almost always of an ulcer on the lip which refuses to heal. For its appearance he has usually a very good reason: it is often attributed to a burn or to an injury. Delay in coming for treatment is often distressingly long. To begin with, the ulcer is small and shallow, but even at the very beginning it has a firm base and an indurated margin which is readily appreciated on feeling the whole thickness of the lip between two fingers. The ulcer becomes raised and increasingly tender to the touch and may bleed and crust over. Secondary infection is often added: œdema, necrosis and sloughing invariably follow. This is especially true of the bulky proliferative nodular type of tumour. Not uncommonly the cancer begins as a deep and often sensitive crack or fissure with indurated margins. A bulky or destructive lesion interferes with drinking and with eating. Drooling, an offensive discharge, distressing halitosis and unremitting pain are features of the late case. At this stage too, anæmia and loss of weight become increasingly obvious.

Diagnosis

It must be taken as axiomatic that any ulcer on the lip of an elderly person which refuses to heal and for which there is no obvious cause, is a cancer. Early resort to biopsy must be the rule. This can always be done without risk under local anæsthesia. It should be adequate and should include a representative portion of the tumour at the spreading edge. A single negative biopsy must never be allowed to over-rule a firm clinical diagnosis. A positive Wassermann should, in most cases, be held to sustain rather than to refute a diagnosis of cancer.

Shallow, indolent and relatively painless ulcers usually covered by a dry crust are not uncommonly met with in the elderly. They simulate cancer very closely.

Differential Diagnosis

A traumatic ulcer typically follows a well remembered injury. It is shallow, very sensitive and seldom indurated. Vertical fissures are, however, often met with in the upper and in the lower lip. The irritation of a pipe stem seems sometimes to be the cause, but more often it is exposure to cold and constant reinfection, which delays healing. The margins of such a chronic fissure may become strikingly indurated and an early biopsy is justified to establish the diagnosis.

The most difficult problem in the differential diagnosis of cancer of the lip is that presented by the degenerative changes seen most prominently in the exposed part of the mucous membrane of the lower lip. Such changes occur most often in the elderly and

lip are equally affected but the commissure is a comparatively uncommon site. The tumour most often begins close to the vermilion border.

Incidence

It is the general experience in this country that the incidence of lip cancer is falling steadily. This is reflected in a slow but steady decline in the number of deaths from this cause each year in England and Wales. The total for 1950 was 161.

Age

It is a disease of the elderly, the mean age in a recent London series being 66 years (Harnett, 1952). It is, however, important to remember that it may be encountered, although infrequently, in the thirties or even in the twenties.

Sex

Males are predominantly affected. They outnumber females in most series by about 10 to 1. Of the 161 deaths from lip cancer in England and Wales in 1950 only thirteen were women. It is, however, a curious fact that women suffer proportionately more from cancer of the upper lip than men.

Ætiology

The incidence of lip cancer is highest in those who follow an occupation in the open air: e.g. agricultural workers, fishermen and bricklayers. It is a rare lesion among the professional classes or in the "well-to-do." The determining ætiological factor is probably exposure to the actinic rays of the sun. Clinical experience suggests that it often occurs in patients with fair colouring and thin weather-beaten skin. The higher incidence in the lower lip can best be explained by the fact that it is more directly exposed to the sun's rays than is the upper. The mucous membrane adjoining a lip cancer often shows changes which experience has taught us to consider pre-cancerous. It may be pale and dry and slightly thickened: alternatively it is thin and cracked, crusted or even superficially ulcerated. Often there is frank leucoplakia, the lip being covered with a thick greyish/white layer showing characteristically a distinctive mosaic pattern. As in all situations where pre-cancerous changes are found, multiple (multicentric) cancers are by no means infrequent.

The cause of these pre-cancerous changes is not always clear. The importance of syphilis in this respect has probably been over-emphasized. Certainly the percentage of patients with a positive Wassermann reaction is seldom significantly higher than in a control group. The case against tobacco is a little stronger. Pipe-smoking is always regarded with a special significance but whether one should blame the mechanical or thermal effect of a pipe stem or some carcinogenic action of the tobacco is not established. The reported high incidence in certain groups of Oriental fishermen whose lips become contaminated with tar, has excited much interest among oncologists.

Pathology

The tumour is in most instances a well-differentiated epidermoid carcinoma of relatively slow growth. It spreads directly to involve the whole thickness of the lip and may pass the commissure to extend to the other lip. In neglected cases complete

(*d*) When properly controlled radiotherapy is not available.

(*e*) When a small early lesion offers an easy alternative to irradiation.

Limited surgical excision invites recurrence. To be on the safe side a clear margin of 2 cm. of apparently healthy tissue on every side is essential. It is wise to monitor the resection by submitting representative blocks of tissue at the margin for frozen section.

It is wise to regard the excision and the repair as two quite separate problems. To restrict the excision in order to facilitate closure is to court disaster. The principles of reconstruction of the resulting defects are considered elsewhere.

When bone is involved, it must be sacrificed. It may be possible to preserve the continuity of the mandible by retaining its dense lower margin. Closure by primary suture can be achieved by advancing the mucous membrane of the floor of the mouth and suturing it to the posterior aspect of the reconstructed lower lip.

B. Of the Regional Lymph Nodes

(1) When there is no detectable enlargement of the regional lymph nodes most clinicians adopt a waiting policy. This implies a complete follow-up system. Less than 10 per cent of these patients can be expected later to develop node involvement: at the first hint of enlargement, a surgical resection is carried out.

(2) When the nodes are enlarged, appropriate treatment is started immediately. It is never allowable to defer resection for long in the belief that the node enlargement is due to infection.

(*a*) *When the node enlargement is unilateral* the choice will rest between:

(1) A supra-hyoid block dissection.

(2) A total block dissection. This is preferred by some surgeons on the score of completeness and is obligatory when nodes in the cervical chain are involved.

(*b*) *When the node enlargement is bilateral*, the usual practice is to carry out a total resection on the worst affected side and a suprahyoid dissection on the other. Alternatively, if only the proximal nodes on each side are affected, the dissection may be limited to the supra-hyoid level on both sides. A staged bilateral total neck dissection is an allowable procedure: a simultaneous ablation of both sides at one sitting is a formidable operation. It may sometimes be possible to remove the primary and the regional lymph nodes in continuity.

(*c*) When the nodes are judged to be inoperable, treatment is usually restricted to palliative irradiation.

Prognosis

The over-all 5-year survival figure is in the region of 60 per cent. If late cases are excluded, it will be nearer 75 per cent. When the tumour measures less than 2·0 cm. across, 80 per cent of patients may be cured.

The outlook for cases which recur after inadequate initial treatment is much less favourable. Involvement of the regional nodes reduces the patient's chances to one-third.

There is little risk of recurrence after a period of 5 years. Any reappearance after this interval is most often due to the development of a new primary.

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especially in men who have spent a great deal of time working out of doors. The lip loses its redness and softness and becomes whiter, firmer and drier. Often it is covered by thin dry crusts which can be removed to uncover a shallow ulcer, which may bleed a little but promptly crusts over again. Such a lesion is precancerous and the patient must, for this reason, be kept continuously under review. There is every justification in such cases for a wide excision of the affected area, advancing the cut edge of the mucous membrane on the gingival aspect of the lip to the skin to form a new vermilion border.

Although the lip is the commonest site for an extra-genital primary chancre, it is a rare lesion in this country nowadays. The central part of the upper lip is the commonest site. A short history, much œdema and prominent rubbery enlargement of the regional nodes may suggest the correct diagnosis. Dark-ground examination of a smear from the ulcer will confirm it.

Retention cysts of the mucous glands of the lip are common. They may be firm, but are usually distinctly blue, and rounded, and are covered by an intact covering of normal mucous membrane. A mixed salivary gland tumour is rare and the history of enlargement extends back for many years. It takes the form usually of a firm rounded lump in the substance of the lip.

A growth appearing at the commissure is very often found to be the anterior extension of a buccal cancer. It is important to bear this in mind. Such lesions are very much more malignant than is a true lip cancer and merit much more active treatment to the primary as well as to the regional nodes.

Treatment

A. Of the Primary

This is one of the most favourable forms of cancer, always provided treatment is given promptly and adequately. Failure to cure is most often due to persistence of cancer at the primary site. Whichever method of treatment is used the patient's fate may well be determined by its thoroughness at the very outset.

There are two alternative methods of treatment: viz. irradiation and surgery.

(1) **Irradiation.** Irradiation in some form is preferred in most clinics. No extensive operation is necessary and the resulting cosmetic and functional result is often excellent.

Cancer of the lip is relatively radio-resistant. It is usually necessary to give treatment in high dosage.

A variety of techniques is applicable, the choice depending less on the type of tumour than on the established practice in the unit.

(*a*) For the average, moderately extensive lesion, high voltage therapy is the rule, with protective screening of the mouth. Teleradium is a useful alternative for the very bulky tumour.

(*b*) Interstitial treatment using a single or double plane implant of radium-loaded platinum needles is equally effective.

(*c*) For a lesion which does not penetrate too deeply into the gingivo-labial sulcus radium mounted in a mould can be applied externally.

(2) **Surgery.** Surgery will be preferred:

(*a*) When the cancer fails to respond to irradiation.

(*b*) When it recurs in a treated area.

(*c*) When it involves bone.

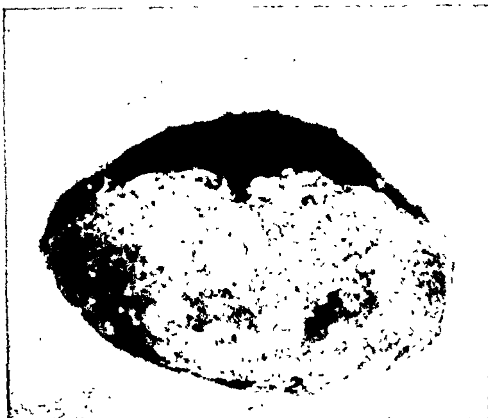


FIG. 97 Macroglossia due to a diffuse cavernous lymphangioma occurring in a child who had had many attacks of infection.



FIG 98. A ranula in a young woman in early adult life

Lane-Claypon, J. E. (1929) Report on Cancer of the Lip, Tongue and Skin. Reports on Public Health and Medical Subjects, No. 59. H.M. Stationery Office, 1929.
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DISEASES OF THE TONGUE

CONGENITAL ANOMALIES

Tongue-tie

This is the commonest of the congenital anomalies of the tongue. In former days its division was a common enough operation in any outpatient clinic. Very occasionally a severe degree of tethering seems to cause feeding difficulties in early life and this is made the justification for a division of the frenulum. More often a reluctant surgeon divides it in infancy when he finds himself unable with confidence to deny that it may be retarding speech development.

Macroglossia

The tongue is one of the more common sites of a diffuse cavernous lymphangioma and this is the most frequent cause of macroglossia. The condition may be noticed soon after birth, and if it is considerable, there will be nursing difficulties. Fortunately the posterior third of the tongue seems to escape and acute respiratory difficulty is unusual. As is the case with lymphangiomatous malformations at other sites infection is a common complication. In a few it will be followed by fibrosis and spontaneous cure, but more often it is a prelude to frequent relapse. The tongue becomes acutely painful and swollen but pus formation does not occur. With each succeeding attack the tongue becomes bigger and thicker and the mouth, teeth, and jaw will become distorted. Often the tongue at this stage is studded with innumerable clear vesicles which are quite distinctive (Fig. 97)

On the assumption that the organism responsible for the infection is streptococcal it is customary to treat the episodes of infection with penicillin. Radiation as a method of treatment is quite unpredictable in its effect. A much more satisfactory method is to control the bulk of the tongue by a V-excision of the central part of its anterior two-thirds.

Ranula

This term is applied to a cystic lesion on the under aspect of the tongue which perhaps belongs more properly to the floor of the mouth. Although well known to all students and illustrated in all undergraduate textbooks, it is in practice a rare condition.

The patient presents with a soft cystic swelling close to the mid-line, covered only by a transparent mucous membrane, across which course some tortuous blood vessels. It is said to have a distinctive bluish-colour and transilluminates (Fig. 98).

The condition is believed to be due to blockage of one or other of the mucus-secreting glands in the floor of the mouth and spontaneous rupture and subsequent refilling of the cyst is a common experience. It can be treated either by excision or by marsupialization.

The tongue is fixed below to the mandible and to the hyoid bone. To remove it completely, the line of section must pass from the base of the vallecula to the floor of the mouth not far behind the gingiva. Many so-called total glossectomies fall far short of this requirement.

When considering the anatomy of the tongue it is important to remember that at the junction of its middle and posterior thirds (and this is a common site of lingual cancer) its lateral margin lies in the closest proximity to the posterior limit of the floor of the mouth, the lower alveolus (and the upper too), the anterior pillar, the soft palate and the tonsil.

It is also apparent that the mucous membrane of the infra-lingual aspect of the free part of the tongue is directly continuous with that lining the floor of the mouth.

Age and Sex Incidence

These have already been considered in an earlier section (pp. 212 and 233).

Site. The different parts of the tongue are not involved with equal frequency. The middle of the dorsum immediately anterior to the sulcus terminalis is curiously immune: it is most unusual to see a cancer beginning primarily at this point. The anterior two-thirds is involved three times as frequently as the posterior third (Harnett). Cancer develops most commonly on the lateral border 5–10 mm. in front of the anterior pillar: but the tip, the lateral margin of the free part of the tongue or its infra-lingual aspect are also common points of origin (Fig. 99).

PATHOLOGY

Cancer of the tongue is in most cases a squamous carcinoma with varying degrees of differentiation.

Anaplastic tumours are encountered most frequently in the posterior third and it is in this vicinity too that the lympho-epithelioma is occasionally encountered.

By and large, the rate of growth of the primary is inversely proportional to the degree of differentiation.

Squamous carcinoma of the tongue often develops on the basis of pre-existing pre-cancerous lesion of which leucoplakia is the most common (Fig. 100). It is not surprising, therefore, that in the tongue as in the corresponding lesions of skin, multiple tumours are relatively frequent. Unless, however, they are widely separated in time, in situation or in their histology, it is quite impossible to differentiate a new primary from a recurrence.

The tumour spreads first widely in the tongue, more deeply than laterally, and soon invades adjoining tissues including ultimately the lower jaw and the cartilages of the larynx. Examination of sections taken right across the tongue at autopsy suggest that it can be taken as axiomatic that the size of a malignant ulcer in the tongue gives little hint of the extent of its invasion in depth.

Involvement of the regional lymph nodes occurs relatively early in the average case. Usually the spread is in a regular sequence, and "skipping" of the most proximal nodes is an infrequent happening. The oft-quoted cases of spread from the tip of the tongue directly to the jugulo-omohyoid node must be a rare occurrence. A well-differentiated tumour may, however, be late in spreading to the regional nodes: and conversely, a small anaplastic primary at the base may at the outset be completely dwarfed by a massive rapidly-growing regional lymph node enlargement.

BENIGN TUMOURS

These are considered shortly under the differential diagnosis of cancer of the tongue.

CANCER

Anatomy

There are certain points of surgical anatomy which are essential to the proper understanding of the pathology and treatment of lingual cancer.

For descriptive purposes the tongue can be considered to consist of two portions which are embryologically and morphologically quite distinct.

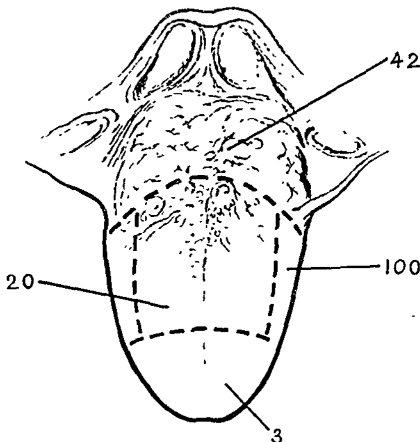


FIG. 99. The relative frequency of cancer in the tongue at different sites (after Richards).

The anterior two-thirds looks mainly upwards and is the only part of the tongue which we see when a patient opens his mouth and pushes his tongue out as far as he can.

It is important to recognize that the anterior faucial pillar marks the posterior limit of this portion of the tongue and that the posterior third or base really belongs to the oropharynx, of which it helps to form the anterior wall. It looks mainly backwards and can be inspected only with the aid of a mirror. It is, however, accessible to the finger, if only for a moment, in even the least co-operative of patients.

The anterior two-thirds is covered with papillæ which vary considerably in their appearance. Under the mucous membrane of the posterior third lie the aggregations of lymphoid tissue which constitute the lingual tonsil.

increased and as swallowing becomes progressively more difficult the constant drooling of saliva is added to his misery. A complete distaste for food may develop relatively early and loss of weight and progressive anæmia invariably follows. The unhappy state of the patient with advanced disease is indeed pitiable, as ulceration, infection and infiltration pursue their relentless course.

Long before this stage the patient has usually noticed a swelling in the neck on one or on both sides and in just over 1 per cent of cases this may be the first symptom.

Posterior Third

When the primary is on the posterior third the symptoms are often less urgent, and the first complaint may be of dysphagia or alternatively of a vague feeling of a lump in the throat. A small but important group (15 per cent of the total) first seeks advice with a painless and often bulky neck swelling below the angle of the jaw which is often bilateral and usually asymmetrical. There are lymph node metastases in 75 per cent of cases (50 per cent for anterior two-thirds). In this latter group it is important to remember that symptoms relating to the primary are often most misleadingly absent.

Findings on Physical Examination

The early lesion on the tongue may present in a variety of ways.

Most frequently it is when first seen an oval ulcer with a slightly raised nodular margin and a pale, red or brownish base which bleeds fairly readily. Alternatively, it takes the form of a sessile warty tumour, the surface of which may be crumbling, bleeding and ulcerated.

Less often it presents as a deeply recessed ulcer which really forms the face of a deep cleft or fissure in a tongue which is often extensively scarred by syphilis.

A well-differentiated slow-growing tumour (again often in a shrunken syphilitic tongue) may infiltrate the tongue widely, until it is represented by no more than a hard, wrinkled, lobulated and fixed structure with little or no surface ulceration (Fig. 101).

Most baffling of all is the lesion, commonly in the posterior third, which may, at least in its early stages, appear as a smooth lump with an intact mucous covering. This is the cancer which is so easy to overlook on mirror examination.

The cancer may show as a warty or ulcerated elevation in the centre of a patch of leucoplakia. alternatively, the epithelial changes typical of this disease will be found elsewhere in the lingual or in the buccal mucosa.

In the female it is worth while looking especially for the degenerative changes in the oral mucosa which go with sideropænic dysphagia and which include a smooth red tongue, and a small oral aperture (Fig. 102).

Quite the most significant feature of all malignant tumours of the tongue is the presence of induration as detected by the palpating finger. This is especially true of the less accessible lesions when infiltration of the pterygoids has led to such a degree of trismus that adequate examination of the whole tongue becomes well-nigh impossible. The examination of a cancer at any site in the mouth is not complete until it has been freely and carefully explored by the gloved finger. As a diagnostic manœuvre in the detection of non-ulcerated tumours in the posterior third, it is of prime importance and it is equally valuable in the investigation of any lesion to which a nervous patient denies ready visual access. The induration may involve not only the obviously ulcerated area, but the

One is seldom impressed clinically by the existence of distant metastases, but deposits widely in viscera and in bones are often found at autopsy. This is especially the case where the primary tumour has, for some time at least, been adequately controlled by treatment.



FIG. 100 Section taken from a patch of leukoplakia on the tongue The changes evident in the epidermis can often be seen evolving into those of a squamous cell carcinoma.

SYMPTOMS

Anterior Two-thirds

When the cancer is on the anterior two-thirds of the tongue, usually the first and main symptom is of a painful lump or ulcer. The pain is, at the outset, felt in the tongue itself and is aggravated by the taking of hot or cold fluids. Later it may be referred (presumably along the lingual) to the area of sensory distribution of the auriculo-temporal nerve to the ear and to the side of the head. Typically, the old man with a lingual cancer, already has a pledget of wool in the auditory meatus when he first comes seeking advice.

Not infrequently pain is absent at the outset and the patient notices only a steadily enlarging sore on his tongue, which obstinately refuses to heal. The wearing of a denture soon becomes impossible but discarding it brings no relief.

Uncommonly a patient first presents complaining of a recent and obvious change in a long-standing patch of leukoplakia, which has become, for the first time, hard, ulcerating, bulky or painful.

Whatever the onset, pain is a prominent feature of the advancing disease. A bulky and necrotic tumour makes eating and speaking difficult, and may bleed often and freely: a more infiltrating lesion is equally disabling. The breath becomes fœtid. Salivation is

tongue widely and diffusely in the immediate vicinity: in estimating the local extension of the disease into adjacent tissues, the evidence which comes from palpation is quite the most informative.

In approximately one-third of cases the cancer is still confined to the primary site: in the remainder there is clinical evidence of regional lymph node metastases. Most often



FIG 103 A typical example of the painless smooth and sessile tumours which are found on the free part of the tongue and which consist of squamous epithelium covering a connective tissue core

those in the sub-mandibular region or in the upper part of the jugular chain (sub-digastric node) are first involved. Later, enlarged nodes will be encountered at a lower level in the jugular chain and occasionally nodes may first be detected at this level. In the early stages a strictly unilateral tumour on the anterior two-thirds will give rise to unilateral metastases When the primary is a large one there is considerable risk of bilateral spread, and this is also a feature of median tumours and of the less well-differentiated ones in the posterior third. Typically, the regional lymph node metastases from the latter are bulky, rapidly growing and seldom hard, and tend clinically to overshadow what may in any event be a relatively small primary. In the average case, however, the

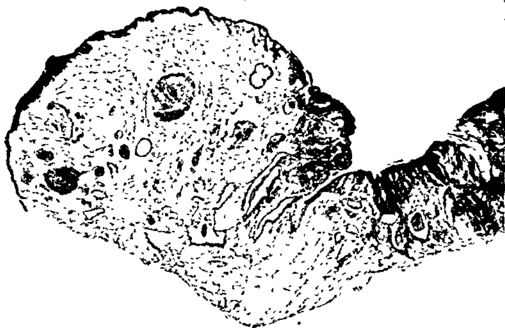


Fig. 101. Section taken from a hard, shrunken tongue which was widely infiltrated by a well-differentiated squamous cell carcinoma with little surface ulceration (×7).



Fig. 102. The typical appearance of the mouth in the established

(3) **Leukoplakia.** The mosaic of thickened whitish epithelium in the classical case is easy enough to recognize, especially if similar patches are to be seen elsewhere in the mouth (Fig. 104). They should be looked for on the buccal mucosa opposite the cutting edges of the teeth, on the palate and on the floor of the mouth. The thickened epithelium looks like a layer of dirty-white and cracked paint · in the milder cases it resembles rather as Butlin described it—"a thin film of boiled white of egg."



FIG 105. Leukoplakia of tongue The "bald" areas near the margin were vivid red in colour

Leukoplakia affects mainly the anterior two-thirds of the tongue and sometimes the centre and sometimes the margins seem to be most seriously affected. It seems highly likely that the pattern changes from time to time as areas of the thickened keratin layer separate off to leave a vivid red smooth patch devoid of papillæ (Fig. 105).

Occasionally leukoplakia is restricted to one part of the tongue and may then present as a localized hypertrophied patch which is diagnosed clinically as either a squamous papilloma or a well-differentiated carcinoma (Fig. 106).

nodes are firm or even hard, discrete and not tender. Later they become increasingly fixed and ultimately tethered to the overlying skin. When the primary is bulky and infected necrosis in the neck nodes is a common sequel: the overlying skin becomes red and stretched, and there presents clinically what looks not unlike a low grade abscess, which the unwary may incise with unhappy results.



FIG 104. The typical patchy white mosaic of leukoplakia. The Wassermann reaction was positive.

Differential Diagnosis

(1) Simple Papilloma. This commonly presents as a slow-growing painless warty lesion on the free part of the tongue often near its tip. It may measure as much as 2 or 3 cm. across. It is usually a sessile proliferative type of tumour, which neither bleeds nor ulcerates and in which there is no hint of induration. It may, however, be impossible other than by a biopsy to distinguish it from a squamous carcinoma of the warty type.

(2) Polyp. Fig. 103 illustrates what is a not uncommon benign tumour often found in the relatively young near the tip. Smooth, sessile, painless and quite soft, its appearance seldom suggests other than a benign diagnosis. Its histology is in no way distinctive. There is a connecting tissue core and it is covered by normal looking squamous epithelioma. Some pathologists like to call it a fibroma.

(5) *In the cancerophobe the self-detection of the prominences of the lingual tonsil or of the lingual folds* (Fig. 107) often causes considerable concern. The facility with which such a patient can demonstrate his entire tongue for examination often gives one a pretty shrewd hint of the correct diagnosis. The congenitally fissured (scrotal) tongue may similarly cause alarm.



Fig. 107. The prominent lingual folds on the posterior margin of the free part of the tongue which patients occasionally mistake for cancer

(6) *Glossitis rhombica mediana* is an obscure, uncommon and painless lesion which takes the form of a red, raw, slightly raised but soft, smooth rhomboid elevation in the midline of the tongue in its middle third. It occurs at a site not often involved by cancer. Its ætiology is obscure and it disappears without treatment.

(7) Syphilis. The tertiary manifestations of syphilis are, in this country at least, now quite uncommon. A gumma is, in fact, a clinical rarity. Typically it is situated in the midline of the tongue somewhere in front of the V-line (an uncommon site for cancer) and presents as a painless lump, the surface of which forms the base of a punched-out brownish sloughing ulcer. Induration is absent.

The distorted tongue showing coarse lobulation is no more common: it is important to recognize that cancer alone, in the absence of syphilis, may present as a deep indurated cleft.

(8) A cancer in the posterior third causing dysphagia must be differentiated from a primary lesion of the oro- or hypo-pharynx.

(9) A small primary in the less accessible parts of the tongue must always be borne in mind as a cause of an enlargement of the sub-mandibular or of the upper jugular lymph nodes.

The change from leukoplakia to cancer may show itself clinically in a variety of ways. Most often a small firm nodule develops in one area, rises up above the surrounding mucous membrane and later breaks down to present as a typical indurated ulcer. Alternatively, one of the cracks between two adjoining patches deepens and widens to leave a hard and painful cleft.



FIG 106 Photograph showing an especially exuberant patch of leukoplakia on the left side of the tongue and the more classical appearance on its right margin The main tumour mass was excised and section proved it to be benign Wassermann reaction was positive

(4) **Traumatic Ulcer.** This diagnosis can be allowed only when the position of a tongue lesion can be shown without question to correspond exactly to some source of irritation of which a ragged tooth edge or a broken filling are the most common. Further, its correction must lead within two or three days to prompt relief from symptoms and to signs of healing.

A pyogenic granuloma presents as a non-indurated ulcerated elevation often near the tip of the tongue, usually at the site of a penetrating wound of which a bite is the most common. Snipping it off with scissors may be enough to encourage prompt healing.

(6) When the whole tongue is diffusely involved by cancer, especially if, as occasionally happens, the tumour is a well-differentiated one and there are no regional lymph node metastases.

In arguing the case for or against surgery or irradiation it must be assumed that adequate forms of each are available. The results of ill-planned treatment of either sort are disastrous. When surgery is used in an attempt to make good the failures of irradiation the results are never happy. Dissection and definition of structures and of tissue planes is difficult, and the recognition of post-irradiation scarring from persisting or recurring tumour impossible. Healing tends to be long delayed and morbidity is high. Radiation in the support of failed surgery has, by and large, less unfavourable consequences.

There has been evident in recent years, a revival of interest in the place of surgery in the control of mouth cancer in general, and partial and complete tongue resections are being practised with an increasing frequency. Improvements in methods of anæsthesia and of blood transfusion and the availability of antibiotic agents, has removed much of the hazard and of the morbidity of this type of work.

The following general surgical methods are applicable, the actual technique involved varying in some degree from one patient to the next.

(1) For a tumour in the free part of the tongue which has not encroached on to the floor of the mouth, liberal excision with a margin of healthy tongue presents no difficulties. Endotracheal anæsthesia, with a pharyngeal pack and a relaxant drug affords easy access. There is no need to do a preliminary lingual artery ligation: the divided vessel can be secured with precision as the tongue is incised. A clean dissection with the cold blade has much to commend it rather than the use of the diathermy needle. The tongue is reconstituted by suturing mucous membrane to mucous membrane or by drawing the tip of the tongue round to fill a gap on one side (Fig. 108).

(2) For a lesion further back on the margin of the tongue and for any lesion involving the floor of the mouth adequate access can be achieved only by turning back a cheek flap and dividing the mandible. Since this dissection involves venturing into the submandibular region, it seems reasonable to do a simultaneous radical neck dissection. The extent of the operation is determined entirely by the size of the tumour as determined at operation: there is no place, for example, for a planned hemi-glossectomy.

Every effort should be made to achieve primary healing. To allow free approximation of the mucous lining it is usually necessary to resect rather less than half the mandible. What remains of the tongue can then be swung across to fill the gap. It is true that this may narrow the oro-pharynx, tether the tongue and by obliterating the sulcus make the subsequent wearing of a denture rather unlikely, but these disadvantages are more than outweighed by the inestimable benefit of primary healing. The mucosal suture line must be supported by a succession of buried sutures approximating such muscular layers as are available.

The airway in the first few post-operative days is assured by a tracheostomy and nutrition is maintained by nasal feeding tube.

(3) Total glossectomy can be carried out if required. It involves only a natural extension of the procedure described above: to fill the gap it is necessary to suture the epiglottis to the floor of the mouth in front.

(4) When the cancer has encroached on the epiglottis it will usually prove necessary

Prevention

The cause of leucoplakia being unknown, there is no satisfactory treatment. It would seem reasonable, if the Wassermann reaction is positive, to give the appropriate antiluetic treatment. Equally, scrupulous mouth hygiene and the complete cessation of smoking would seem to be worth enforcing. A localized patch of leucoplakia can occasionally be excised completely by a simple surgical excision. Most often, however, its proper management consists of an unremitting follow-up at regular intervals with the most careful visual and digital exploration of the whole tongue. The slightest hint of any change is the signal for an immediate and, if negative, a repeated biopsy.

A female patient with a smooth tongue, anæmia and dysphagia can be in some measure protected against the risk of cancer in the tongue by the adequate and continuous administration of iron in an assimilable form. Of the merit of the administration over prolonged periods of a preparation of crude yeast to patients who have raw, red tongues, as a prophylactic against cancer, there is no firm proof.

Treatment

Although there are obvious disadvantages in considering separately the management of the primary and of the regional lymph nodes metastases, since many of the problems relating to the latter are the same whatever the site of the primary growth, we propose to consider the whole problem of metastatic cervical cancer together in a later chapter: here we are dealing mainly with the treatment of the local disease in the tongue.

Curative. *Although in former times lingual cancer was treated surgically, its management has, in this country at least, now become mainly the concern of the radiotherapist. Even if the wisdom of this change is debatable, there are certain cases which seem eminently suited to some form of radiation therapy.*

(1) Cancer of the posterior third (which is in general not readily amenable to surgical excision) especially if it is poorly differentiated and associated with bilateral lymph node metastases.

(2) When the patient refuses operation, or is unfit for surgery, on account of his poor general condition. Age is, of itself, no deterrent to surgery: even the very old stand the widest resections extraordinarily well. It must also be remembered that there is a morbidity associated with radiation treatment which is not inconsiderable. Desquamation of the mucous membrane in the treated area and lessening of the salivary flow makes eating and swallowing difficult and loss of appetite and a distaste for food are not infrequent.

Surgical treatment is by many confined to the following circumstances:

(1) Lesions confined to the free anterior portion of the tongue, the more so if they are bulky.

(2) When there is involvement of bone.

(3) When the tumour has failed to respond to irradiation.

(4) When there has been a recurrence in an area which has already been given a dose of irradiation up to the limit of tolerance.

(5) When there is a coincident leucoplakia and especially if the tongue is scarred by syphilis. The non-cancerous adjoining portions of the tongue under such circumstances do not readily tolerate the required dosage of irradiation and tissue necrosis is a common sequel.

invites the occurrence of painful necrosis and the tedious and painful discharge of the sequestrated bone which follows.

The hazard of over-irradiation is always present, even when the dosage is carefully assessed and controlled. Tissue necrosis, painful and persistent ulceration and considerable disability are then the rule. Differentiation of radium necrosis from a persistence or recurrence of the tumour under these circumstances is exceedingly difficult. cancer is not uncommonly overlooked in the belief that ulceration in the treated area is due to the effect of irradiation.

Lingual cancers vary widely in their responsiveness to radiation therapy. In general, the less well-differentiated tumours show the most striking and heartening immediate response, but radiosensitivity and radiocurability seldom in this situation go hand in hand. By and large, the chance of cure is highest in the group where differentiation is most apparent, but as in cancer at other sites, the degree of responsiveness to radiation can never be forecast with any great precision.

Palliative. More than one out of every ten patients coming for treatment with lingual cancer will be considered at the very outset to be too advanced for treatment.

There can be little merit in wide resections, which are known to be incomplete: quick recurrence is the invariable result. The crude diathermy excision of the main part of the tumour in the mouth is a method which has its adherents.

A great deal can, however, be done to mitigate the sufferings of his latter days by:

(*a*) Scrupulous mouth hygiene.

(*b*) By controlling super-added infection by the administration of the appropriate antibiotic.

(*c*) By the institution of tube feeding.

(*d*) By nerve section.

Earlier this used to be achieved by division of the lingual nerve in the mouth but this is seldom adequate. Alcohol injection of the mandibular division of the trigeminal or division of its main sensory root, are more likely to be effective.

Radiotherapy has little to offer as a method of palliation of the primary growth: painful ulceration is an invariable sequel.

Prognosis

The natural history of the disease untreated is 12–18 months, death from bronchopneumonia, lung abscess, inanition or urgent bleeding coming mercifully to bring an end to their suffering

Although the prognosis is considerably affected by whether or not there are lymph node metastases (52 per cent as against 15 per cent 5-year survivals—Radiumhemmet) it is important to remember that the majority of patients at death still have cancer persisting locally in the tongue: the real difficulty in treatment is control of the cancer at the primary site.

Butlin in 1909 reported the results of the surgical treatment of 197 cases of lingual cancer. No less than fifty-five of these was surviving at 3 years which, even if we admit that this was a selected group of relatively early cases compares favourably with present-day figures from the major radiotherapy clinics. Figures for modern surgical treatment are not yet freely available, but it is quite apparent that even the widest resections can now be practised with a low mortality and morbidity.

to sacrifice the larynx. It may still be possible to make good the large resulting defect on the anterior wall in such a way as to achieve primary healing.

Radiotherapeutic technique is now very highly specialized: there is no longer any place for the surgeon who, without the guidance of a physicist, disposes radium needles interstitially in the tongue.



FIG. 108. The appearance of the tongue 7 months after excision of approximately one-third of the free part of the tongue There was just perceptible speech impairment.

There is no standardization of practice. The method in vogue varies widely from one clinic to the next. The objective of all methods is the same, namely, the selective destruction of the cancer cells, with the minimum damage to the adjoining healthy tissues. Some prefer teleradium or X-rays from an external source: others interstitial radiation or a combination of the two methods. Diathermy may be used to excise any persisting nodule in the tongue at the end of treatment. Extraction of all teeth in the irradiated area would seem to be a wise precaution before embarking on treatment, as would the adequate control of dental caries elsewhere in the mouth. Some prefer a complete dental clearance as a routine preliminary. There must be no dental manipulations in the treated area for years thereafter. A dental extraction or any operation on this bone immediately

figures that cancer of the mouth can no longer be considered a common disease. For example, even in the average large metropolitan teaching hospital new cases of cancer of the tongue will be seen at the rate of only 2 per month (say 2 per cent of all cancer cases) so that, in this country at least, the average general surgeon is unlikely, even in a lifetime, to acquire any considerable experience in its management.

INCIDENCE ACCORDING TO SITE

The tongue is the commonest site of cancer in the mouth, followed by the tonsil, the cheek and the gingiva. (The order of frequency varies somewhat in different series.) The palate is the least often affected.

It is, in fact, often a matter of guesswork to say exactly where a tumour has begun—whether, for example, primarily in the tongue, or in the vallecula, tonsil, the floor of the mouth or alveolus. When, however, we are intent on comparing methods of treatment at various sites in the mouth, it is important that we make every effort to make as precise and accurate an anatomical diagnosis as possible.

AGE

Cancer of the mouth does not often occur in early adult life: the great majority of cases are seen after the age of 55. The relative youth of a patient is, however, never by itself a valid reason for the exclusion of cancer from the list of differential diagnoses.

SEX

Cancer of the mouth is almost four times as common in men as in women. This male preponderance is noted at all sites, but is less apparent in the younger patients.

It is interesting that the steady decline in the number of deaths each year (as shown, for example, in the figures for cancer of the tongue) affects mainly the figures for males, so that the percentage incidence among women is rising steadily each year (Fig. 109). It would seem apparent that this change is due to the diminishing effect of some ætiological factor which operates predominantly in males. In contrast, the significantly high figure (45 per cent) for the incidence among women in Sweden must point to the operation in that country of some factor peculiar to the female sex (see below).

SOCIAL INCIDENCE

Although there is no obvious relationship between occupation and the incidence of buccal cancer, the incidence of the disease rises as one descends in the social scale. It is encountered more often in hospital than in private practice, but this is less apparent today than it was 15–20 years ago.

CAUSATIVE FACTORS

Leucoplakia

Of the ætiological significance of leucoplakia in the causation of cancer of the mouth there is general agreement. The close relationship between the two has been demonstrated in two ways:

(1) If all patients with leucoplakia are followed closely, a significantly high percentage will be found to go on to development of cancer in the affected area. Since the interval

Of patients treated by radiation roughly 20–30 per cent will survive for 5 years but the outlook is only about half as favourable in patients who have concomitant syphilis or leukoplakia.

The prognosis varies in relation to the size of the primary and the site of the tumour also affects the outlook very considerably. The cure rate for cancer at the base is less than half that for tumours on the anterior two-thirds.

CANCER OF THE ORAL CAVITY

INCIDENCE

In 1950 in England and Wales, out of a total population of 44 millions, the Registrar-General reported approximately 1,300 deaths from cancer of the mouth (including the oropharynx). Comparable figures for stomach, breast and rectum were 14,000, 8,000 and



FIG. 109 Graph showing the steady fall in the number of deaths from cancer of the tongue in *males* between 1931 and 1950 (From Registrar-General's Statistical Review, 1950.)

6,000 respectively. If one assumes that the number of deaths each year from this disease is a true index of its incidence, there has been a steady decline in its frequency during the past 20 years. This is true for cancer at all sites in the mouth and is shown dramatically by the figures for lingual cancer (see Fig. 109). It seems unlikely that more effective treatment is alone responsible for this improvement. It will be evident from these

thermal or mechanical effects of a cigarette or of a pipe-stem is strongly suspect as a carcinogenic agent. There seems to be little doubt that betel-nut chewing* in certain communities in the Orient leads to cancer of the cheek, but constituents of the quid other than tobacco are usually held responsible. The case against tobacco in this country is by no means a strong one for, if it was important one might reasonably expect to find a rising curve in its incidence to match the increasing prevalence of the smoking habit (as is abundantly so in the case of lung cancer). Instead, as we have seen, the incidence of the disease is falling steadily.

Degenerative Changes in the Mucous Membrane

There is, both in Sweden and in Finland, a relatively high incidence among women of cancer of the mouth. There is little question that this frequency is directly related to the prevalence in Scandinavian women of atrophic changes in the buccal mucosa, which are noted in about half the women with buccal cancer. They are exactly similar to those observed in the tongue and pharynx of patients with the Plummer-Vinson syndrome, with which we are more familiar in this country as a predisposing cause of post-cricoid cancer in women. It seems clear that the conditions are identical; certainly sideropænia and dysphagia are common features of each.

The important clinical aspect of this association between degenerative changes in the mucosa and cancer is the realization that the latter is in some degree preventable if the anæmia is vigorously treated from the outset by adequate dosage of iron in a readily assimilable form.

It is widely believed in the United States that similar degenerative changes in the mucous membrane of the tongue and elsewhere in the mouth are attributable to deficient intake of one or other or several of the Vitamin B fractions and that they too predispose to cancer. The taking of crude yeast in heavy doses is claimed to lessen this risk.

It is highly probable that the absorption of iron and of vitamin B are, in fact, closely inter-related.

PATHOLOGY

Cancer of the MOUTH is predominantly squamous cell carcinoma showing varying degrees of differentiation.

Metastases are in the first instance to the regional cervical lymph nodes. Multiple primaries are by no means infrequent.

In the tonsillar area there occurs, but with some rarity, the lymphoepithelioma, a tumour in which there is a curious blending of epithelial elements and of lymphoid tissue. Willis holds the view that the tumour is essentially a poorly differentiated squamous epithelioma, and that the incorporation of lymphoid tissue can reasonably be explained by the close proximity in certain situations of epithelial and of lymphoid cells.

CLINICAL FEATURES

The patient usually presents with an ulcer which will not heal, or alternatively with an ulcerating tumour. Pain becomes a prominent feature in the later stages, as does fœtor of the breath and progressive weight loss.

There are certain distinctive features of tumours at different sites.

* Sanghvi, L. D. RAO, K.C.M., Khanolkar, V. R. *Brit. Med. J.* 1955, 1, 1111.

between the first appearance of leucoplakia and the development of cancer may be as long as 15–20 years, any worth-while follow-up must be a very long one. It is, no doubt, for this reason that carefully documented studies of this type are by no means common and the evidence on this score is based more on clinical impression than on fully recorded experience.

(2) By noting in what a high percentage of patients with buccal cancer, leucoplakia is present in the adjoining mucous membrane, whether it be in the obvious case detected clinically or in a lesser degree when noted later by the pathologist in routine sections.

The relationship between leucoplakia and cancer is most apparent in the tongue. For example, Harnett (1952) reported leucoplakia in 33 per cent of all cancers on the anterior two-thirds, but in only 10 per cent of those at the base. It will be seen later that there is a significant parallel between these figures and those both for the distribution of cancer and for the frequency of a positive serological test for syphilis in the same areas.

As to the cause of leucoplakia we can only guess, but there seems to be little doubt that there is not one but many and that of these syphilis is, at least in these modern days, no longer the most important. Further, it is important to remember that the close relationship between leucoplakia and cancer of the tongue is noted whether the Wassermann reaction is positive or not.

Syphilis

Although syphilis is commonly held to be an ætiological factor of some moment in cancer of the mouth at all sites, it is only in the tongue that there is any really convincing evidence of its importance in this respect. Among patients with lingual cancer the percentage with a positive serological test is significantly higher than that among a comparable group taken from the same population. Further, that syphilis is especially significant as an ætiological factor in relation to cancer of the anterior portion of the tongue seems clear from the fact that the percentage of cases where a positive Wassermann reaction is recorded is four times as high in cancer of the anterior two-thirds of the tongue as it is in cancer of the base.

It is unlikely that syphilis, *per se*, is a precancerous condition: cancer does not develop in a gumma. It is, however, of considerable importance, firstly by being at least in part responsible for leucoplakia and secondly, by leading occasionally to the development of widespread scarring (especially in the tongue) which similarly seems to predispose to cancer.

The ætiological importance of syphilis in oral cancer is much less evident today than it was 10–20 years ago and the declining frequency of the tertiary manifestations of this disease may well be reflected by a corresponding progressive fall in the incidence of cancer.

Oral Sepsis

Although often held to be of major importance in the causation of lingual cancer, no one has shown convincingly that broken, mis-shapen, carious or filled teeth, or dentures however worn or ill-fitting, are in any way responsible for cancer of the tongue. If they ever lead directly to the development of cancer, they certainly do so but rarely.

Tobacco

Tobacco, whether it be from the presence of some chemical in the smoke or from the

kind of tumour from a primary lip cancer which carries a much better prognosis. It does not take long for a tumour to penetrate through the whole thickness of the cheek (Fig. 110) and the ultimate disfigurement may be hideous. When the primary is far posteriorly the metastases may first be evident in the nodes which lie in front of the ear and in close relationship to the parotid gland.

Although, strictly speaking, cancer of the tonsil belongs to the pharynx rather than to the mouth here, as at other sites, it is often exceedingly difficult to know the precise point of origin of the tumour. Early lymph node enlargement is a prominent feature of cancer of the tonsil and a swelling in the neck is not uncommonly the first sign. The primary may be so small and so silent as to be completely overlooked. The primary tumour will, at a relatively early stage, spread through and beyond the superior constrictor to involve the closely related great vessels which will ultimately become hopelessly implicated.

DIAGNOSIS

. It must always be borne in mind that in the elderly the commonest cause of a chronic progressive and localized ulcer or tumour anywhere in the mouth is cancer: equally, however, this diagnosis must be entertained earlier in life for cancer does occur, though rarely, in the twenties and thirties.

The patient is almost invariably able to account for its presence by some very simple explanation, be it a tobacco or food burn, a bite, pressure from a ragged tooth or a worn denture. In general, the physician would be wise to reject every such explanation out of hand at the very outset and to reconsider it only if there is incontrovertible evidence of its validity. Of all the clinical features of a malignant ulcer, induration is quite the most significant.

Speed in diagnosis is a matter of the greatest importance. A blood Wassermann reaction should be taken as a routine but a positive result, if anything, supports a clinical diagnosis of cancer. It is seldom justifiable in a doubtful case, either to await the outcome of a course of anti-luetic treatment, when a gumma is suspected, or the effect of the removal of a ragged tooth when a traumatic cause is held responsible.

BIOPSY

It has never been shown that the taking of a biopsy increases the risk of rapid dissemination or prejudices the chance of ultimate recovery. The clinician should never have the slightest hesitation in its use. To turn to biopsy at the very outset is not an indication of clinical inexperience and of diagnostic ineptitude: it can be a sign of mature judgement.

To be of value the biopsy must be in every sense adequate. Most helpful is a deep biopsy taken through the growing edge. It is an advantage to take several fragments from different portions of a tumour, especially if it is a necrotic growth. Like most diagnostic methods, biopsy has its limitations. If the clinical diagnosis is cancer, never rest content with a single negative biopsy. Take a second, a third, or even a fourth. Equally, however, it is often impossible without a biopsy to establish the diagnosis of cancer. This is especially so when one suspects a recurrence in a treated area. Time is often lost and all too frequently with fatal consequences, by attributing continuing ulceration to the effect of treatment by radiation. Where a tumour recurs years after apparent cure by radiotherapy biopsy is especially necessary to establish the diagnosis, although the academic question as to whether the cancer is a persistence of an original

A cancer beginning in the alveolar mucosa will later spread medially to the floor of the mouth and to the tongue and latterly to the lip or cheek. There may, at a relatively early stage, be radiological evidence of erosion of bone and ultimately there will be gross destruction with direct extension of the tumour through the entire thickness of the overlying skin and the development of a fixed fungating ulcer. When situated far posteriorly

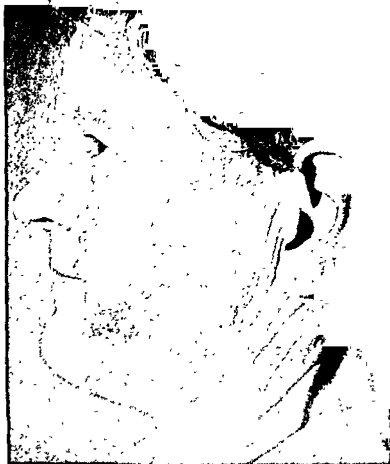


FIG 110 Carcinoma of the mucous lining of the buccal cavity which had penetrated the entire thickness of the cheek.

it will spread quite soon to the posterior part of the floor of the mouth, the side of the tongue and the anterior pillar Experience proves that there is in such cases often a hidden extension into the region of the pterygoid muscles.

Cancer involving the floor of the mouth soon presents as a swelling below the jaw. This is much more often due to direct extension of the primary rather than to lymph node metastases but both methods of spread may be in evidence together. In a neglected case an offensive salivary fistula is almost invariable.

Cancer of the buccal mucosa begins most often opposite the line of closure of the teeth and is not infrequently based on a diffuse leukoplakia. Its anterior margins will ultimately involve the commissure of the mouth and it is important to separate off this

Tonsil

A cancer of the tonsil is always to the patient in the early stages no more than a "sore throat": delay in diagnosis is almost always the rule, the primary is often small and difficult to see and attention may be distracted by a large deposit in the regional lymph nodes.



Fig. 111. Prosthetic epulis of 12 months' duration, resulting from ill-fitting dentures. Female, aged 32 years. There was no ulceration

TREATMENT

In general in this country the treatment of cancer of the mouth has come to be primarily the concern of the radiotherapist. The proportion of cases dealt with surgically is in most centres very small indeed. It tends to be offered only as a second best where there is failure of response to irradiation.

The hazard of intensive irradiation *near bone* is necrosis with slow and painful separation of sequestra, so that surgery has come to be preferred as the method of choice for cancer in this situation. It is true that surgical excision presents considerable technical problems, but cancer of the mouth at most sites can be excised with little danger and with a reasonable chance of having acceptable cosmetic and functional results. The hazard of

one or due to a new primary or to the undoubted carcinogenic action of irradiation is usually insoluble.

DIFFERENTIAL DIAGNOSIS

All Sites

In establishing the diagnosis of cancer at various sites in the mouth the following points should be borne in mind. A circumscribed patch of leukoplakia may resemble a cancer very closely and an adequate biopsy will decide the issue.

Similarly a squamous papilloma may be indistinguishable from a slowly-growing cancer: it should, if possible, be dealt with by excision biopsy.

With the manifestations of lichen planus on the mucous membrane the dermatologist is well familiar. Occasionally patients come first to a surgical clinic and it is well to bear in mind the small greyish-white and slightly elevated lesions of this skin disease as they present in the mucous membrane of the mouth. They are smooth and velvety and resemble cancer not at all—not uncommonly, however, they are incorrectly called leukoplakia with all the implications that follow such a diagnosis.

Floor of the Mouth

A large submandibulary salivary calculus ulcerating through into the mouth is a rarity.

Gingiva

Any adult whose denture for no very obvious reason ceases to fit properly should be suspected of having a gingival (or palatal) cancer. It is true that mucosal hypertrophy does sometimes occur as a result of the chronic irritation of a badly fitting denture but it is not ulcerated and should not give rise to much difficulty in diagnosis. It usually presents as an elongated, fleshy and often furrowed ridge on the outer aspect of the alveolus (Fig. 111). On the hard palate a similar lesion may develop in relation to the central part of the denture as a flat papillomatous lesion which is again non-ulcerated.

A fungating bleeding tumour which fills an empty socket after dental extraction for toothache not infrequently turns out to be a gingival cancer, or in the case of the upper jaw, an antral cancer erupting into the mouth. An obvious part of the investigation of any gingival tumour is an X-ray of the underlying bone. The intra-alveolar epidermoid cancer arising in the jaw and only later erupting into the mouth (Willis) although rare, is worth bearing in mind.

Most of the epulides are granulomatous in nature and some are non-ulcerated.

Palate

The majority of malignant ulcers on the hard palate originate in the antrum: it is often quite impossible to decide whether they have begun in the mouth or in the mucosa of the antrum. A complete clinical and radiological investigation of the superior maxilla and of the nose is required in every case.

The palate is one of the commoner sites of a gumma but it is typically a central, painless, ulcerating lesion with no heaping up of its edges and no induration.

Mixed salivary tumours of the palate are very slowly growing and rounded and ulcerate only in the later stages.

BENIGN TUMOURS

Fibro-Osseous Tumours

This is a group of tumours which consist of varying amounts of fibrous tissue and of bone and which involve not only the upper and the lower jaw but the other bones of the face and skull. The condition may be localized or diffuse and can, by its behaviour, perhaps best be regarded as a dysplasia of bone rather than a true neoplasm. The clinical



FIG. 112. Congenital Epulis The tumour was successfully removed in the first days of life. (Mr Selwyn Taylor's case)

presentation varies a good deal from one patient to the next and pathologists take a great delight in dividing them up on the basis of subtle changes in histology, into a host of different types, each of which is given its own distinctive name.

We can, however, for all practical purposes, consider them all together as an ill-understood group of tumour masses associated with bone which are probably due to abnormalities in its normal development. They are first seen most frequently before the

this type of operation is immensely less with modern endotracheal anæsthesia and the free use of the antibiotics. Post-operative morbidity is much reduced by primary closure. In the fashioning of lining and of skin flaps great help can be had from the plastic surgeon but elaborate staged restorative procedures have no place until the risk of local recurrence has passed. Resection of the primary can readily be combined with a simultaneous *en bloc* neck dissection.

TUMOURS OF THE JAW

EPULIS

Congenital Epulis

The term epulis is not a very satisfactory one. It implies no more than a tumour on the gum. This is a rare tumour of the gums of new-born infants; it presents as a firm rounded non-ulcerated tumour usually on the pre-maxilla. Although some of the cells of these tumours show cross striations, the presence of which has earned for them the title "granular myoblastoma," there seems to be some doubt about their muscular nature. It is probable that it is a hamartoma rather than a true tumour.

The specimen illustrated in Fig. 112 was pedunculated and was removed without difficulty and did not recur.

Epulis of Pregnancy

This seems in most cases to be no more than a hypertrophic gingivitis due to deficient mouth hygiene and to dental sepsis, which becomes more active and more obvious during pregnancy. Its control is entirely a dental problem.

Giant-cell Epulis

This is the commonest variety. It is found most often in early adult life as a firm smooth dark red swelling on the outer (labial) aspect of the jaw, commonly in the lower jaw and somewhere in front of the pre-molar teeth. In the great majority of cases the tumour arises in relation to teeth but is seen occasionally in the edentulous jaw. The condition is painless There is no radiological abnormality in the underlying jaw.

On section the tumour may be found to contain very many giant cells and to resemble closely an osteoclastoma To this the name "giant-cell" or "myeloid" epulis is sometimes given but there is good reason to believe that the great majority of epulides of this type are perfectly benign and that they are most probably infective rather than neoplastic in nature.

It is enough to remove the tumour surgically and to excise the remaining teeth.

Multiple Myelomata

Rarely an epulis is found to be a plasmocytoma and may be the first (and even for a time, the only) manifestation of multiple myelomata

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age of 20 and do not increase in size after the end of the growth period. They have none of the aggressive characteristics of a true neoplasm.

The condition may involve the jaw and the bones of the face generally: if it is widespread and symmetrical, the clinical diagnosis is leontiasis ossea. Alternatively, a single bone or only part of a bone may be affected.

Clinical Features

The condition is usually noticed in adolescence when a painless swelling of the bone gradually becomes apparent. It takes the form of a hard, slightly lumpy and ill-defined swelling, usually involving the outer aspect of the bone. Frequently it is a small lesion affecting only the alveolus or the hard palate.

Radiologically it is marked by an area of dense sclerosis but when it contains a lot of fibrous tissue there will be no more than mottling of the bone.

Differential Diagnosis

Sarcoma often suggests itself as a possible diagnosis when a young person presents with a bone tumour. It is, however, a rare condition which is both painful and destructive.

Paget's disease affecting the bones of the face may be radiologically very similar, but this is a disease of older people.

Treatment

A small tumour can easily be removed with a chisel. It is not necessary to excise it completely: a rough trimming procedure is all that is necessary. Operation has the great merit of allowing the diagnosis to be established beyond dispute.

If an entire bone, or many bones are involved, a planned operation to correct gross facial asymmetries should be postponed until the growing period is over.

TUMOURS OF DENTAL ORIGIN

Peri-apical Granuloma

Chronic infection of a tooth root leads to destruction of the adjoining bone. This cavity becomes lined with granulations which are found clinging to the root of the tooth when it is extracted.

Dental Cyst

Dental cysts occur in connection with the permanent teeth and are seen more often in the upper than in the lower jaw (Fig. 113). It presents as a hard swelling, which expands the outer plate of the alveolus. It may become so attenuated that it gives the "lubricating-can" crackle. In the upper jaw it may present on the hard palate or grow upwards into the antrum. It usually occurs in relation to a dead or infected tooth. Its pathogenesis is a matter for dispute but the view now widely held is that in most cases it follows infection and may be no more than nature's effort to line with epithelium a chronic cavity in bone.

Dental cysts are treated by extracting the associated tooth and then removing widely the top of the cyst, followed by excising as far as is possible its epithelial lining. The remaining cavity is then packed and is allowed to close gradually from below. In treating a large dental cyst of the maxilla it may be necessary to remove a considerable part of the floor of the antrum.

Dentigerous Cyst

Dentigerous cysts are thought to arise from the enamel organ which forms a cap over the normal developing tooth. Should the enamel organ become cystic the tooth is prevented from erupting and is left with a crown projecting through the wall of the cyst (Lucas).



FIG. 113. Dental cyst in an edentulous jaw.

Its site is marked to begin with by an unerupted tooth, followed by a swelling with expansion and thinning of the bone of the alveolus.

Radiologically there will be a *translucent area with a clear-cut edge, and projecting* into it the crown of an imperfectly developed tooth which is often widely separated from its proper site of development.

Treatment. A dentigerous cyst is removed in precisely the same way as a dental cyst taking care, however, to extract along with it the related tooth.

Adamantinoma

This is a rare but distinctive tumour, which although locally destructive, does not metastasize and which is characterized by a very long natural history.

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tumour-like malformations." They result from some deviation of the normal development of the tooth-forming element in the jaw, mesodermal as well as ectodermal.

Clinically it presents as a swelling of the jaw, usually in early life, often associated with pain and with infection as it endeavours to erupt. One or more teeth are found to be missing at the site of the tumour.

Radiologically it is recognized by the density of the shadow it casts. This investigation usually rules out the alternative clinical diagnosis of an unerupted or supernumerary tooth.

An odontome should be removed as soon as it is diagnosed to minimize the distortion of the remaining teeth. This may involve cutting away a covering layer of bone to give access to the tumour which can then be loosened and elevated. There is no question of recurrence.

MALIGNANT TUMOURS

Squamous Cell Carcinoma

The commonest malignant tumour of the jaw is that which follows its direct invasion by a tumour on the adjoining mucous membranes or skin. A growth on the alveolus will involve the underlying bone at an early stage and this is also true of a neglected cancer of the floor of the mouth or of the lip, or of the antrum. The gross destruction of bone in the recurring rodent ulcer is fortunately less frequently seen than it was in former times.

Squamous cell carcinoma may, however, begin in the bone of the jaw. This is recognized most frequently in the mandible but it may equally well occur in the maxilla. Willis believes that it takes origin from remains of the dental lamina, the ridge of epithelial cells which join together the enamel organ and the ectoderm of the mouth. It invades the bone widely and ulcerates secondarily through into the mouth often in several places. Its true nature tends to be overlooked by the clinician, who may treat by surgery or by radiation, only that portion of the tumour which presents in the mouth. It metastasizes in exactly the same way as does a squamous carcinoma of the gingiva.

Metastatic Carcinoma

The bones of the jaw are not often involved by metastases from other sites. It is, however, seen occasionally in patients with widely disseminated cancer of the breast or lung and also from primaries in the kidney, thyroid or adrenal medulla.

Osteogenic Sarcoma

The jaw—upper or lower—is a very uncommon site for this tumour. Pain and swelling are early and persistent symptoms and there will eventually be severe limitation of function. The tumour tends to be rapidly progressive. Often there is redness and swelling of the overlying skin or mucous membrane. Radiologically there is evidence of bony destruction: the classical signs of new bone formation as seen in sarcoma of a long bone are seldom evident.

A firm diagnosis is made only after an adequate biopsy has been carefully examined at leisure.

The outlook is always unfavourable. It tends to run a rapid course with early and wide dissemination There seems little to choose in an early case between a wide resection

Clinical Features. It is a tumour of early adult life, most patients presenting themselves for treatment in their early thirties or forties. The only complaint is of a slowly progressive facial disfigurement. Since the condition is, in the great majority of cases, completely painless, the tumour may already have been present for over five or ten years. Occasionally a tumour is first spotted by the dentist when he is fitting a new prosthesis.

Although it may well be that we often overlook an adamantinoma originating in the upper jaw and bulging upwards into the antrum, it is certainly true that in the recorded cases tumours in the mandible are at least four times as common as those in the maxilla.

The tumour begins in the centre of the bone in the region of the last molar tooth and as it grows it destroys and expands the bone, but almost always at the expense of the outer wall. A smooth firm swelling becomes apparent in the region of the angle of the jaw.

As it grows it usually extends upwards into the anterior part of the ascending ramus and palpation of the thinned and expanded bone may give the characteristic "eggshell" or "lubricating-can" crackle. A neglected tumour will grow to immense proportions and involve the entire jaw. Although the teeth may loosen and fall out, the stretched and distorted mucous lining of the mouth remains intact and ulceration or bleeding are very uncommon symptoms.

Pathology. Although the histological picture is a variable one, the characteristic features are a pallisade of tall cylindrical cells abutting on a connective tissue stroma and containing a cyst or a collection of fine stellate reticular tissue.

Its pathogenesis is still hotly debated but the case for its origin from the epithelial cells of Malassez is argued with most persuasion.

For all practical purposes the tumour remains only locally destructive: distant metastases do not occur.

Diagnosis. The long history of the steady development of a painless swelling in the region of the angle of the jaw in a person in early adult life usually suggests the diagnosis. The clinical features of a mixed parotid tumour are not dissimilar but careful physical examination and an X-ray will rule out the presence of a lesion in the bone.

Although the radiological appearance may be diagnostic it is very variable and can be simulated very closely by a dentigerous or a dental cyst. In the classical case there is a multiloculated destructive lesion of the jaw with sharp scalloped margins.

A firm diagnosis is essential as a preliminary to treatment, based on a generous biopsy, blocked in paraffin and examined at leisure.

Treatment. The older conservative operations, which consisted for the most part in simple curettage, were followed by recurrence which was almost invariable. Even a radical resection gives a promise of success in only three out of every four cases. It is necessary to turn back a cheek flap and to remove the whole tumour, and at least 1·5 cm. of adjacent healthy bone. In the average case the functional and cosmetic result of such an operation is quite acceptable. The fitting of a graft or of a splint is often quite unnecessary at the primary operation and a later reconstruction is required in only a minority of cases.

Although some confident claims have been made for the use of radiotherapy in the cure of adamantinoma surgical opinion has certainly not yet been won over to its favour.

Odontomes

Few subjects in surgical practice are so confused by a complicated nomenclature as are the odontomes. They are exceedingly rare Willis described them as "benign

Diagnosis. The diagnosis is very easy to overlook. Pain in the face, toothache, an ill-fitting denture, epistaxis, nasal obstruction are all common enough symptoms; but the development of any of them in the elderly without an obvious cause should immediately suggest cancer of the maxillary antrum as a possible diagnosis.

An X-ray will show an opacity of the antrum and later destruction of its boundary walls.

The only way to be certain of the diagnosis in many cases is by taking a formal biopsy of the antrum after exploring its cavity through the canine fossa or by removal of a tissue fragment if the tumour is already presenting in the mouth or nose or on the cheek.

Treatment. In most clinics the favoured treatment is by a combination of surgery and radiation.

The surgeon first explores the antrum to confirm the diagnosis and establishes free drainage into the nose (after removal of a large portion of the inferior turbinate). The tumour is then irradiated, either by a standard 250 k.v. machine or by a teleradium. Six weeks later the antrum is explored through the mouth removing the greater part of the floor of the antrum and of the adjacent nasal cavity. The bony walls of the antrum along with any residual tumour are now as widely removed as is possible and the defect in the palate and alveolus is closed by an obturator. Through this large window the cavity can be inspected for the early detection of recurrences which can then be treated by intracavity radium.

Lymph node involvement is treated either by a block dissection or by including the upper part of the jugular chain in the area of irradiation.

Increasing attention has been given within recent years to the possibilities of more radical surgery in carcinoma of the antrum. The Weber-Fergusson technique involves turning back a large cheek flap and excision of the entire maxilla, including also the floor of the orbit (and the eye itself, if the tumour has infiltrated in this direction). If the cheek flap is lined with a Thiersch graft healing occurs quite quickly. The resulting deformity is inconsiderable if a dental plate can be fitted early in convalescence with a supporting mould to prevent collapse of the cheek flap.

Prognosis. Of the patients who are given treatment which is aimed at cure, 1 in 5 might be expected to be surviving at the end of 5 years.

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of the bone and radiotherapy. The latter is used as a palliative when the tumour is inoperable.

Osteoclastoma

The jaw is an uncommon site for a giant-cell tumour. Clinically there is a tumour of the jaw—most often the mandible—which expands and thins the cortex and presents in the mouth as a dark red swelling. Radiologically there is a defect in the bone, often crossed by septa dividing it into a number of loculi. It will resemble closely an adamantinoma. Usually the diagnosis is established only by biopsy. The giant cell tumours of hyperparathyroidism are histologically identical so it would seem wise in all cases to estimate the serum calcium level. Treatment will be by surgical excision. Strong claims have been made for the value of radiotherapy in these cases. Radiologically the area of bone destruction appears to increase in the early stages of treatment, but this is followed later in a successful case by progressive healing with recalcification.

Cancer of the Antrum

The commonest malignant tumour of the upper jaw is a cancer, originating from the mucous lining of the antrum but it is often quite impossible to decide whether a tumour comes primarily from the antrum or whether it has spread there from the alveolus, the hard palate, the nose or the ethmoids.

It is met with most commonly over the age of 50 and in most series men outnumber women.

Clinical Features. A tumour arising from the mucous lining of the antrum remains hidden until it has filled up the antral cavity and has begun to extend beyond its bony walls. Pain is, however, a prominent symptom and is typically much more persistent than it is in cases of uncomplicated antral suppuration.

When it grows from the lower and most medial part of the antrum it may show itself relatively early by erupting through the palate or alveolus into the mouth. The patient comes complaining that his denture no longer fits, or alternatively he has severe toothache and may be found to have a loose tooth. Its removal affords no more relief than does the extraction of the adjoining sound ones: on the contrary, the pain may become more severe and the empty socket bleeds in a most troublesome way and is seen to fill up with soft greyish tissue which in no way resembles healthy granulations. Eventually there will be a fistula from the antrum into the mouth. Should the tumour erupt forwards it will cause a progressive swelling of the face with obliteration of the nasolabial fold or a prominence at the inner canthus.

Extension medially through the thin lateral wall of the nose will be followed by unilateral epistaxis and nasal discharge or by the development of polypi, infected or malignant. The latter bleed very readily and are seldom quite translucent.

A tumour which erupts upwards will give rise to neuralgic pain by involvement of the inferior orbital nerve with anæsthesia (or less often hyperæsthesia) of the skin of the cheek. An extensive tumour will displace the globe of the eye and cause diplopia or even proptosis. Blockage of the naso-lacrimal duct shows itself clinically by epiphora.

Enlargement of the regional lymph nodes, especially those in the sub-mandibular triangle and in the upper deep cervical chain is noted in almost one-fifth of all cases at the time when they first come under treatment.

artery, to the internal jugular vein. There are several large inferior thyroid veins passing from the inferior parts of the lobes to the innominate veins. Uncommonly an additional artery, the *thyroidea ima*, enters the inferior surface of the isthmus; it arises from one or other of the larger arteries in the neighbourhood.

Other important relationships are the recurrent laryngeal nerves and the parathyroid glandules. The latter, usually two in relation to each lobe, are variably sited but each superior one is commonly behind a superior pole, each inferior one near the entrance of the inferior artery or behind or below the inferior pole. They lie most commonly loose on the thyroid surface; sometimes they are embedded in the surface. The two recurrent laryngeal nerves pursue a slightly different course. The left, given off by the vagus in front of the aorta, bends under the aorta to ascend in a more median plane than the vagus along the line of the anterior border of the œsophagus but inclining forward to reach finally the groove between trachea and œsophagus and thus to have a posterior-internal relationship to the lateral lobe. The right nerve issues from the vagus in front of the right subclavian artery and loops under and behind the latter in an outward direction so that it ascends from a more lateral position than its fellow, passing at first behind the start of the common carotid artery to reach its inner side. It then passes up behind the lateral lobe to enter the larynx, as does its fellow, by piercing the cricothyroid muscle just posterior to the prominent cricothyroid articulation. In its ascent each nerve may pass in front of, behind, or between the entering branches of the inferior thyroid artery. The nerve may sometimes be kept closely applied to the lobe surface by surrounding fascia; in others it may be somewhat embedded in the surface.

The thyroid receives branches from both the sympathetic and parasympathetic divisions of the autonomic system. Its lymphatics gather into a plexus on its surface whence there is drainage upwards to glands in front of the larynx, laterally to the deep cervical glands, and downwards to pretracheal glands.

The gland is made up of innumerable microscopic somewhat spheroid units, the follicles, which are usually lined by a single layer of cubical epithelium. In the lumina of these there is storage of colloid. The state, size and contents of the follicles varies greatly in disease.

Physiology. The secretion of the thyroid, the thyroid hormone, is essential for maintaining the normal level of metabolism in the body; additionally it is essential in the young for normal growth and development. Absence or atrophy of the thyroid in the developing fœtus leads to cretinism, the widespread changes, particularly mental, of which are but little affected by the administration of thyroid extract. Atrophy starting in childhood leads to juvenile myxœdema which must be recognized and treated early by the extract if irreparable damage is not to be sustained by brain and body. Atrophy in adult life leads to a myxœdema which yields readily to regular medication with thyroid extract.

The thyroid has relations with other ductless glands and their inter-related physiology is a complex affair as yet far from completely unravelled. But it would seem that in the main the thyroid is activated by the thyrotropic (thyroid stimulating) hormone of the anterior pituitary. This thyrotropic hormone, provided there is an adequate supply of iodine available in the body, stimulates the thyroid gland to produce its own hormone, the thyroid hormone, which is discharged into the blood stream to stimulate the metabolism of the tissue cells throughout the body. Additionally, the released thyroid hormone acts upon the anterior pituitary itself in such a way that the amount of thyrotropic hormone

CHAPTER V

THE THYROID: AND NECK IN CHILDREN

C. DONALD and DAVID EVANS

ANATOMY AND PHYSIOLOGY

Anatomy. The thyroid consists of two lateral lobes, applied to the anterolateral aspects of the lower part of the thyroid cartilage, cricoid, and upper trachea and pharynx. An isthmus, with its upper margin just below the cricoid, commonly connects the two lobes. The isthmus may be absent; the whole gland then has an almost completely anterior relation to the trachea. At the other extreme, the isthmus may be wide and connect two deeply set lobes. The latter form is not uncommon in men and knowledge of it is important as such lobes may be considerably enlarged without a goitre being obvious, well-developed overlying muscles aiding the concealment. Intermediate between the extremes is the common type with a small square isthmus which, if enlargement of both lobes occurs, is soon obliterated. Each lateral lobe normally measures about 1½ in. vertically, about 1 in. at its widest, and about ¾ in. at its thickest. Each tapers upwards to a superior pole; the rounded lower end is called the inferior pole. A smaller narrow lobe, the pyramidal lobe, runs upwards from the isthmus towards one or other side of the thyroid cartilage, usually the left. It is a remnant of the thyroglossal tract and is of some importance in that, if not removed at operations for toxic goitre, it may hypertrophy and reproduce the disease. It is absent in some 20–30 per cent of thyroids. Each lateral lobe has the infrahyoid muscles closely applied to it anterolaterally; deeper than this the carotid sheath is a lateral relation.

The blood supply is abundant and comes from four main vessels and a number of unnamed fine vessels derived from tracheal and other arteries. These latter open up widely when the gland is over-active and then supply more than enough blood to the remnants of the gland when it has been subtotally resected and all the main vessels divided or ligated. Each superior thyroid artery, derived from the external carotid, comes down on its superior pole. It does not, however, enter the apex, but after giving off a branch to the posterior aspect of the lobe at this point, courses down in front of the pole before dividing into branches which enter the gland. Since the emerging superior veins, going to the internal jugular, have the same relationship, this is a point of operative importance. Their division at operation some distance below the apex of the lobe allows the latter to be prised gently out of the upper parts of the neck when the lobe is much enlarged; otherwise there may be great difficulty in applying ligatures above the apex of the lobe. The inferior artery of each side is a branch of the thyreo-cervical trunk of the subclavian, and passes, always deeply to the common carotid, in a loop of variable size, convexity upwards, to divide into several branches close to the gland, which they enter on the postero-lateral aspect about the junction of the upper two-thirds and lower third. There is no middle thyroid artery; the middle thyroid veins are inconstant in size and number and run directly lateral from the gland, always in front of the common carotid

Lingual Thyroid. The gland may wholly or in part fail to descend with the thyroglossal outgrowth of the ventral wall of the primitive pharynx. It then develops in the tongue, in close relation to the foramen cæcum. It has been asserted that complete failure to descend is about three times commoner than partial failure, when there is thyroid tissue both in the tongue and in the neck; in the latter instance it is either normally situated or lies near the hyoid. Radioactive iodine studies will no doubt in future make these points more accurate since negative palpation alone, the neck only sometimes being explored, is insufficient proof. The reported cases of lingual thyroid, a rare anomaly, show a preponderance of females to males as 7 to 1.

The condition usually remains unnoticed until its enlargement occurs. It is rarely noted before puberty and most reported cases have become obvious by the age of 30. Enlargement may be accompanied by such symptoms and signs as dysphagia, dyspnœa, bleeding and ulceration. Hyperthyroidism is rarely associated; hypothyroidism is a more common feature. The physiological responses at such times as puberty, pregnancy and the menopause are similar to those of the normal gland and naturally lead to more severe symptoms. Similarly there may be the pathological changes of simple goitre with its degenerations. Bleeding may occur at the times of menstruation.

In size the lingual thyroid varies from that of a small grape to an orange; in shape it is most commonly spherical; it may be completely embedded in the tongue or partly embedded, partly projecting from its surface; occasionally it is pedunculated. Its surface is smooth or somewhat roughened; the colour is usually a red-brown; when stretched by a projecting tumour the overlying mucous membrane is thinned and may be ulcerated. The bleeding results from the latter state which may involve large veins lying in the surface of the thyroid tissue. The consistence is variable, depending upon the physiological or pathological states. The cut surfaces of excised lingual goitres show pictures similar to that of normally situated glands. Carcinomatous change has been observed in the male. Associated parathyroid tissue has been demonstrated on histological section.

Benign tumours such as adenoma and lipoma, and malignant tumours such as carcinoma and lymphosarcoma may occupy a similar position. While biopsy will establish a differential diagnosis, the use of radioactive iodine will obviate such need if the tissue be thyroid.

Medical treatment in the form of prolonged iodine therapy has caused recession in size of the gland. Similarly, the permanent ingestion of thyroid extract has led to disuse atrophy. Operative treatment is indicated when obstructive symptoms or bleeding are severe. The operation can usually be performed through the mouth; a preliminary tracheotomy may be required. Where the mass is deep in the tongue it may be better approached from the neck as in the radical operation for a thyroglossal sinus. Complete excision is best lest further swellings or carcinoma supervene. Substitution therapy will be needed later if there is no thyroid tissue in the neck.

Thyroglossal Cysts and Fistulæ. Persistence of the thyroglossal duct, in whole or in part, leads to the development of fistulæ and cysts. The upper opening of the original duct, which embryologically is an outgrowth of entoderm of the ventral wall of the pharyngeal portion of the gut, corresponds eventually to a point in the bottom of the foramen cæcum of the tongue. From this the track passes downwards and forwards in the midline of the substance of the tongue to the front of the middle of the hyoid bone and then for a variable distance beyond this, depending on the degree of caudal displacement

is controlled. Thus a balanced level of circulating thyroid hormone is achieved in health through what has been called "the pituitary thyroid axis." This self-regulating mechanism seems to be necessary for the pituitary and thyroid to cope with altering conditions and stimuli which affect them, presumably mainly from the hypothalamus.

A supply of iodine is necessary for the production of thyroid hormone. Where this is lacking, as in simple goitre, the level of thyroid hormone in the blood falls. This causes stimulation of the thyrotropic hormone but, iodine not being sufficiently available, the effect is hypertrophy and hyperplasia of the thyroid without increase in production of thyroid hormone.

There is a further relationship between anterior pituitary and thyroid, independent of the thyrotropic hormone, connected with growth; this will not be discussed here.

The workshops of the thyroid are its vesicles, the lining cells of which manufacture both the contents of the vesicles, the stored colloid thyroglobulin, and from this as needed the hormone itself. The exact chemical structure and synthesis of both is as yet incompletely known but, in respect of physiological activity, thyroxine is the essential element in both. The colloid, however, can be formed without iodine and requires iodination before the hormone can be produced. Radioactive iodine studies show that iodine normally stays as iodide a few hours in the blood plasma, several weeks in the thyroid gland, and only a few days as thyroxine in the blood stream. Taken up from there by the tissues it re-enters the blood stream as iodide, partly to be excreted by the kidneys, partly to be taken up again by the thyroid.

Variations from the normal can be detected by the introduction of radioactive iodine, which is chemically identical with ordinary iodine, into the blood stream, either intravenously or by absorption from the alimentary canal. The iodide is cleared from the blood stream by the kidneys and the thyroid. Estimations may be made of how much is cleared by each and comparison made with normal standards. Where the thyroid is abnormally active, as in toxic goitre, the clearance by the thyroid becomes greater in proportion to the urinary clearance, so avid is the gland for iodine. In myxœdema a reverse picture is obtained. A drawback to the clinical value of these estimations is that there is some variation in normal standards; the lesser differences of functional activity cannot therefore be accurately demonstrated.

Anomalies of the Thyroid

Anomalies of the thyroid from developmental aberrations may be due to failure of its descent, full or partial, to descent beyond the normal, or to abnormal displacements, resulting in ectopic tissue. Anomalies of the thyroglossal duct also occur.

Failure to descend may result in the gland developing in the tongue or hyoid region. Ectopic thyroid tissue has been found within the pharynx, œsophagus, and trachea, and along the outside of the trachea. Abnormally low descent may result in intrathoracic goitre, although the commonest cause of this is the pathological downward prolongations of normally situated glands. It is almost always by the pathological enlargement of these abnormally situated portions of thyroid tissue, particularly in the form of simple goitre and its degenerations, that they become noted. The tissue may be the only thyroid tissue in the body. Thyroid tissue has also been found in ovarian teratomata.

The chief anomalies which require elaboration are lingual thyroid, thyroglossal cysts and fistulæ, and suprahyoid cysts and fistulæ.



discharge of pus. The fistulous opening thus produced tends to heal and break down intermittently. The infective process may sometimes be much more acute towards a similar termination.

The term "cyst" is loosely applied to the unruptured swelling which is strictly not a cyst since it does not have a complete wall, being breached at one point by the track above. It more resembles a small lake fed by a tiny stream. But there is one variant, quite rare,

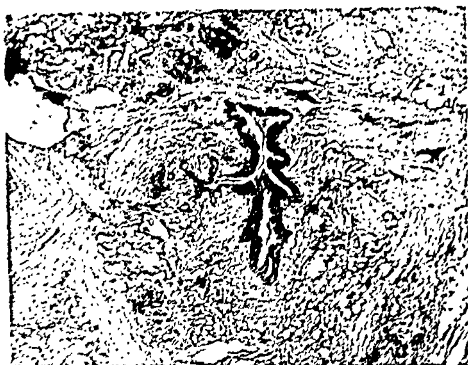


FIG. 116. Thyroglossal fistula. Transverse section of excised fistula in same patient showing diverticula.

which is a true thyroglossal cyst with a complete wall. It would seem to be due to persistence of a small portion of the tract at its lowermost end, growing quite large (Fig. 117) and showing little liability to infection; it can be removed as an unruptured cyst; because of its size it tends to be dislocated to one or other side of the midline, usually the left.

The opening of the ruptured "cyst," the fistula, is essentially midline but may be slightly to one side, previous inadequate attempts at removal may lead to its being even further lateral. When the opening has been present, even intermittently, for several months or more, there develops above it a crescentic fold of skin with its concavity downwards (Fig. 118). This is due to the tethering of the tract to the skin at the opening; at every movement of swallowing the tethered part is pulled upwards and inwards beneath the loose skin above.

The differential diagnosis of thyroglossal tract remnants is from other midline or near-midline neck swellings and fistulæ. Dermoid cysts may occur anywhere along the midline but are chiefly near the hyoid or just above the suprasternal notch. Small ones near the hyoid are quite common and it may be impossible to differentiate these from a thyroglossal cyst. Both are in such intimate relation with the hyoid that they move

of the thyroid gland or upon the size of its pyramidal lobe. Its persistence, from a clinical point of view, is commonest in its most distal part; swellings and sinuses confined to the supra-hyoid portion are infrequent and will be discussed later (p. 256). Demonstration of a complete track by lipiodol is shown in Fig. 115. But it is usually impracticable thus to show the extent of a track before operation for its removal and it is *equally impracticable to identify the length of the* persistent track during operation.



Fig 114 Thyroglossal fistula Boy aged 8. Intermittent discharge for 3 years

It is stated above that the duct usually only persists in its lower part. One reason given for this assertion is that injections of lipiodol into a thyroglossal sinus rarely pass more than a centimetre or so upwards. This, however, is hardly a good reason since the tracts have often been the seat of recurrent infection and undoubtedly at some points are too narrowed and tortuous to accommodate the passage of the thick lipiodol. A better reason is that recurrence of a sinus is uncommon if the track below the hyoid and the middle portion of the hyoid bone are removed. But this is no extenuation of an incomplete operation.

Most of the tracks are therefore sinuses rather than fistulæ. It is better, from the viewpoint of operation, to regard them as fistulæ. The lining epithelium is usually columnar, occasionally squamous.

Clinically, a small swelling appears under the skin in the midline of the neck, most commonly just below the hyoid but also anywhere between the hyoid and the suprasternal notch. The majority appear in patients of between the ages of 3 and 7 years but parents maintain occasionally that the swelling has existed since birth; their first appearance may occur as late as in early middle age. There is commonly a history of a preceding upper respiratory infection or exanthema, it would seem as if the infection lit up the secretory activity of the lining membrane

The swelling, which rarely becomes bigger than a small grape, may remain unruptured for months, fluctuating a little in size and even occasionally disappearing, to reappear later. *In this quiet phase* it contains a glairy substance exactly resembling fresh white of egg.



Fig 115 Thyroglossal fistula Same patient as Fig 114 Lipiodol injection showing the complete track Note the diverticula The prolongations at the top of the track indicate lipiodol spreading over surface of tongue

Infection, however, occurs sooner or later in the contents and is usually of mild degree. The overlying skin becomes tethered tot he swelling, turns slowly red and gradually thins to rupture with the

Treatment. Excision of the complete tract is essential if recurrent fistula formation is to be avoided. Incomplete operations are too frequently performed. If recurrence takes place it is usually within a few weeks or months of incomplete operation; occasionally it happens many years later.

While it is probably true that the track in most instances does not persist for any great distance above the hyoid, and that it will commonly be adequate to remove the track below the hyoid and the middle portion of the hyoid itself, such an operation may be incomplete and, especially because of the hyoid removal, will make further intervention awkward. It is not difficult at the original operation to carry the dissection up to the foramen cæcum.

Determination of the length of the track by the use of probes is a useless procedure; only the subhyoid portion is likely to be traversed. Again, above the level of the hyoid it is unlikely that a persistent track will be seen with the naked eye at operation. Attempts to make it apparent by injections of coloured fluids into the fistulous opening just before operation have a theoretical but not a practical basis; the fluid is apt in the subsequent manipulations to suffuse through the tissues outside the track. It has been recommended that such injections should be made on several occasions at weekly intervals but not for a week before operation; the track wall has then absorbed enough to show up at operation. But this and other bothersome and time-consuming procedures are unnecessary although where there is a fair-sized fistulous opening it is worthwhile injecting lipiodol to obtain an X-ray picture. The wall of the track above the hyoid is of no strength and tears readily; it has to be excised within a wall of muscle. This is best done by "coring out" a somewhat conical portion of the interior of the tongue from the hyoid to the foramen cæcum, not a particularly difficult procedure if a diathermy cutting needle is used.

The technique where a fistula is present is as follows. A general anæsthetic is given and an intratracheal tube introduced. A transverse incision, in or parallel to a skin crease, but embracing the fistulous opening or temporarily healed scar, is mapped out. For purposes of hæmostasis the subcutaneous and subplatysmal layers of the neck are infiltrated with normal saline containing adrenalin to the strength of 1 in 150,000 (6 minims of 1 in 1000 adrenalin to 50 c.c. of saline) over the area in which the flaps will be reflected up and down. The incision is made down to the subplatysmal level and the flaps retracted. The lower flap need only be raised a short distance; the upper flap will require to be lifted in its centre well above the hyoid and thus, especially if the fistulous opening be low, the incision must be several inches in length. Particular care must be taken when reflecting the upper flap not to cut too deeply over the actual line of the track which for practical purposes is subcutaneous below the hyoid. The platysma does not exist in the midline of the neck below the hyoid and it is not difficult, where the opening is low, to cut clean through the tract and raise it with the upper flap. If a probe can be introduced this mistake is easily avoided. The track is then dissected up as far as the hyoid. The scalpel is now laid aside and the diathermy needle employed. The middle portion of the hyoid is now removed. In the first few years of life the hyoid can be cut with a scalpel; thereafter a bone-cutting forceps is required. When baring the bone before cutting, the shape of the body of the hyoid has to be remembered. The dissected tract and the excised hyoid, attached to one another, are now in continuity with the suprahyoid muscles and the tongue musculature. Reference to Fig. 119 will show that the deviation towards the foramen cæcum now alters to about 45 degrees. There is no need to take any more of the

similarly upwards when the tongue is protruded. The tongue protrusion test is only satisfactorily carried out when the patient is asked to open the mouth, protrude the tongue and then warned not to shut the mouth when asked to withdraw the tongue; simultaneous movements of tongue and jaw vitiate the proper observance of the test. Sometimes the swelling is so rounded and palpable between finger and thumb that one can say with fair confidence that it is much more likely to be a dermoid cyst. Tuberculous glands occur occasionally in the suprasternal notch but there are usually other enlarged glands elsewhere; they are alleged also to occur higher in the midline as solitary swellings but



(By courtesy of the Editors of the "Practitioner")
FIG 117 Thyroglossal cyst Girl aged 5

this must be rare Infection of a dermoid cyst may give rise to a transient sinus; the breaking down of a tuberculous gland will give rise to one of much longer duration. Dermoid cysts and tuberculous glands in the suprasternal notch will, of course, not move upwards with protrusion of the tongue.

A small adenoma of the pyramidal lobe of the thyroid in an adult may stimulate a thyroglossal cyst. Only the persistence of the swelling for years without any inflammatory complication is likely to make the diagnosis absolute. More important in this connection is the realization that the whole of the thyroid tissue of the body may very rarely be concentrated in a small rounded mass in front of the hyoid. Before removing any solid thyroid tissue from this region it will be well to ascertain whether there is a gland in the normal situation. Otherwise, permanent myxœdema may follow. Differentiation of a thyroglossal from a branchial fistula is easy. The latter has always a minute opening situated laterally at the anterior border of the sternomastoid muscle. A rare condition, *midline cervical cleft*, may be confused. This is probably due to failure of proper fusion of the branchial masses. There is a vertical, partly raw, partly cicatrical cleft in the midline of the neck, with occasionally a tiny sinus at one point.

Thyroiditis

Acute Thyroiditis: Chronic Specific Thyroiditis: Riedel's Disease: Lymphadenoid Goitre (Hashimoto's Disease): Subacute Pseudotuberculous Thyroiditis

Acute Thyroiditis. Acute infective inflammation of the thyroid most commonly follows upper respiratory infections such as tonsillitis, but may also occur during fevers. While of apparently rare occurrence its real incidence may well be greater, since it often does not proceed to the suppuration which clinches the diagnosis. Some of the painful tender localized swellings which are attributed to hæmorrhage into a cyst may well be inflammatory, because of the rapid disappearance of the swelling.

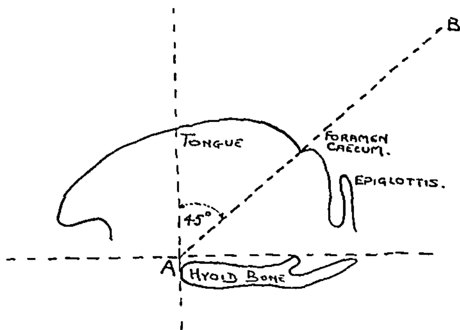


FIG. 119. Sistrunk's operation. Diagram to show direction of the track from the hyoid bone to the foramen cæcum

The inflammation may affect the whole gland or part of it or be limited to a nodule. No ill-effects on the function of the gland have been observed; the condition has not been noted in toxic or malignant goitre.

The whole gland, a lobe, or a nodule swells and is painful. There is a variable degree of malaise and fever. The process may subside spontaneously or go on to suppuration, the pus usually tracking to become subcutaneous low down in the midline of the neck. The bacteriology of such pus is most commonly staphylococcal or streptococcal; in some instances associated with fevers the specific organism has been found. Release of the pus leads usually to rapid relief and recovery of the patient.

In a few instances the infection is severe and much dysphagia is caused. With the close relationship of thyroid to trachea there will be tracheitis and this may be severe enough to lead to a little spitting of blood. Rupture of the abscess into the trachea has been noted.

Where the infection is limited to a nodule the affair may follow the same course, except that a chronic abscess occasionally forms. Hæmorrhage into a cyst leads to the

mylohyoid muscle than the lowermost fibres attached to the excised hyoid; the midline raphe of the mylohyoid is split deeply, as are also the anterior fibres of the geniohyoglossus. The divided margins tend to retract and it is well to prevent this at the deeper levels by means of temporary guy stitches which not only allow of more easy apposition later but permit divided deep vessels to be caught more easily. The anæsthetist is asked to introduce his finger into the patient's mouth and to press the tongue downwards and forwards from the region of the foramen cæcum. This helps the operator with direction and shortens the length of tissue now to be excised. There is no need usually to excise



FIG. 118. Thyroglossal fistula Girl aged 10. Note the crescentic upper margin.

so much that the mouth cavity will be entered on the surface of the tongue; it is sufficient to stop and remove what one has cored out when only a thin partition separates the palpating finger of the operator from that of the anæsthetist. The resulting cavity is then obliterated by catgut sutures. It is unnecessary to bring together the remaining portions of the hyoid bone by wiring as is sometimes advocated; approximation of the adjacent muscles is sufficient. No disability follows from the removal of the central portion of the hyoid. The platysma and skin are then sutured as for any neck wound so as to produce a fine scar. Drainage is not usually necessary.

Supra-hyoid Cysts and Fistulæ. Swellings and sinuses confined to the suprahyoid portion of the thyroglossal tract are uncommon clinical entities. If reference to lingual thyroid swellings (p. 251) is here omitted there are left blind sinuses at the foramen cæcum, which are of little or no importance and are indeed mainly found by diligent autopsy search, and occasional solid or cystic tumours derived from the tract. Reference to Fig. 116 will show that the persistent duct within the tongue is not a simple hollow tube but presents diverticula invading the muscular substance irregularly. The cysts and tumours may show in part on the surface of the tongue. Where they are far back it may be easier to remove them through a neck incision than through the mouth.



FIG 120. Hashimoto's disease. Female 56. Goitre noticed for 3 years Note the hypothyroid facies and diffuse goitre

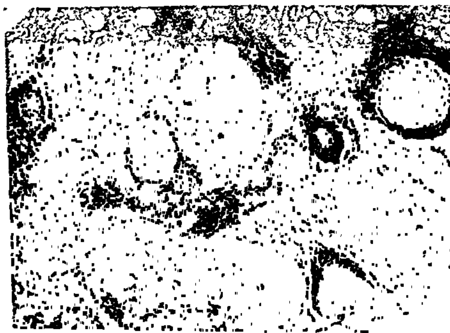


FIG. 121. Hashimoto's disease Microscopic section of thyroid gland removed from female aged 67. In this section five "germ" centres are apparent (×35).

only simulating condition. The two constitute the only examples of painful tender goitres apart from malignant goitre in its late stages. The hæmorrhagic condition has a much more rapid development and is rarely accompanied by constitutional features.

Treatment of acute thyroiditis follows the general line of treatment of infective inflammation by rest, local heat and chemotherapy. Prompt incision is indicated where suppuration has occurred. The rare chronic abscesses cause few symptoms and are usually only met with accidentally in dealing with nodular goitres.

Chronic Specific Thyroiditis. Tuberculous, syphilitic and actinomycotic lesions have been described in the thyroid. They are all exceedingly rare except that *tuberculosis* commonly occurs in the miliary form of the generalized disease; it is but an inconsiderable item in that disease. A focal form is of more interest. Here a hard mass develops in one lobe and gradually softens into an abscess which erupts, if untreated, to the skin surface or into the trachea or œsophagus; secondary pyogenic infection follows. The disease is secondary to tuberculosis elsewhere in the body. Diagnosis is usually made at operation when the affected lobe should be widely resected. Cultures should be taken from any chronic abscess found in the thyroid. *Syphilis* is most commonly seen as a gumma, although a more diffuse enlargement has been noted in the secondary form of the disease. The commoner form, because of its hardness, is easily confused with carcinoma and with Riedel's disease. Diagnosis, if made before operation, should be followed by antisyphilitic treatment which is usually successful. If unsuccessful, operative removal is indicated. *Actinomycosis* occurs as a secondary feature of the disease elsewhere.

Hashimoto's Disease (Lymphadenoid Goitre): Riedel's Disease: Subacute (Pseudotuberculous) Thyroiditis (Crile)

These three goitres are commonly discussed under a comprehensive heading of chronic thyroiditis. This suggests that they are all of inflammatory origin but this is by no means established. Their ætiology, their nature and their inter-relationships are subjects which provoke much argument.

It will be most convenient to describe the three diseases in their most characteristic details and thereafter to discuss exceptional features and to enter into the controversy.

Hashimoto's Disease (Lymphadenoid Goitre). The designation "Hashimoto's Disease" is preferable to that of "lymphadenoid goitre" since the latter suggests a connection with Hodgkin's disease which is alleged by no one. Characteristically (Fig. 120) it occurs in women at, about, or past the menopause who present with a moderately large symmetrical smooth goitre, firm but not hard, and with some degree of hypothyroidism chiefly indicated by increase of weight and slowness of mind. Myxœdema eventually follows. The cut gland at operation is "meaty" and displays no suggestion of colloid storage. Microscopically (Fig. 121) the whole gland is diffusely infiltrated with lymphocytes, here and there interspersed with focal aggregations of the same, some of the latter showing germinal centres; the connective tissue of the gland is greatly increased; the thyroid vesicles are generally small, contain little colloid, and are often atrophic.

Riedel's Disease (Woody or Ligneous Thyroiditis). Riedel's Disease typically appears in men or women at the age of 30 to 45. The goitre is small or moderately large, of almost stony hardness and of smooth surface both lobes are usually but not invariably affected. There are no features of hypo- or hyperthyroidism. The chief complaint is of a feeling of constriction, and severe pressure effects, chiefly on the trachea, may result.



FIG. 120. Hashimoto's disease. Female 56 Goitre noticed for 3 years Note the hypothyroid facies and diffuse goitre.

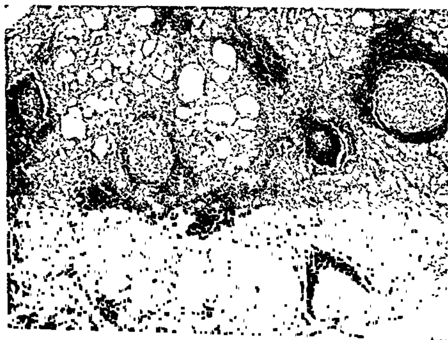


FIG 121. Hashimoto's disease. Microscopic section of thyroid gland removed from female aged 67. In this section five "germ" centres are apparent (×35).

There may be pain in the region of the gland or in the upper parts of the neck. Rarely the main vessels become constricted by the pathological process extending beyond the gland; the recurrent nerve may be similarly involved.

At operation the muscles overlying the thyroid may prove somewhat difficult to separate from it and show by this and by a whitish layer on their deep surface that they have been infiltrated by fibrosis. The gland requires quite forcible cutting, is white or



Fig 122. Hashimoto's disease. Female 39. Excised specimen. Note the pale homogeneous appearance

pinkish white on section, and is much more adherent to the trachea than is normal. The extreme lack of vascularity is a striking feature. Microscopy (Fig 123) shows dense fibrous tissue with little or no trace of remaining thyroid structure; where the sclerosis is less marked, islands of thyroid tissue may be seen surrounded by fibrous tissue, as if the former were being strangled out of existence by the latter. Evidences of hyperplasia of epithelial elements are seen in the less affected portions.

The absence of hypothyroid features in the patient, despite the intense widespread fibrosis of the gland, is strikingly noticeable. It would seem that the body can derive enough thyroid hormone from relatively few functional elements.

It should be interposed here that Riedel, in his original observations, described the sclerosing fibrosis as spreading far beyond the gland proper so that the outline of the latter was lost; there was invasion of adjacent structures and the neck became widely infiltrated with a hard fixed mass which compressed trachea, œsophagus, and carotid sheath. Riedel, on exploring his original case, thought he was dealing with carcinoma; it was its subsequent course which led him to the recognition of the peculiar non-malignant condition.

Few surgeons, even among those who have made a lifelong specialty of thyroid work, have seen the disease in the extreme form described by Riedel. But they have kept applying the term "Riedel's Disease" to a fibrous sclerosis of the gland in which the process has not spread beyond it, or no further than to involve the continuous deep fibres of the infra-hyoid muscles.

Subacute Pseudotuberculous Thyroiditis. This uncommon thyroid affection is characterized by the accompaniment of febrile symptoms and by a histology in which giant-cells are a feature. The presence of the latter has led to the condition being called tuberculous or pseudotuberculous. But tubercle bacilli have never been demonstrated and the giant-cells probably represent a reaction to the escape of colloid from the vesicles (Crile).

The disease tends to follow upon a general malaise. There is, in variable degree, fever, pain and tenderness of the gland. The latter is only slightly enlarged. While the basal metabolic rate is increased the gland takes up much less radioactive iodine than does a normal one. Thus the increased B.M.R. is due to the fever. There is no evidence of reduced thyroid function.

Crile considers the condition to be due to a virus infection. The natural course of the disease is towards spontaneous regression which may occur in a week or two or take several months.



FIG 123 Riedel's disease Female 38 Microscopic section showing thyroid gland replaced by dense fibrous tissue infiltrated slightly with lymphocytes, plasma cells, eosinophil and neutrophil leucocytes. Extremely few thyroid cells left (none in this photograph) Extension of fibrotic process into muscle. No evidence of myxœdema clinically (× 52).

DISCUSSION. There are many examples of "chronic thyroiditis" which do not fall clearly under any of the three titles given above; there has been overmuch tendency to include them all under these. It is likely that in time more varieties will be more exactly defined as to their ætiology and pathology.

It would not seem likely, from the description given above, that there was likely to be any relationship between Riedel's and Hashimoto's diseases. Yet many in the past have regarded, and some still regard, the former as the end result of the latter, the new formation of connective tissue in Hashimoto's disease increasing and increasing until it reaches the dense sclerosis of Riedel's disease. While Hashimoto had specifically stated that he did not hold any relationship between his disease and that described by Riedel, Ewing, on a survey of only 4 cases, maintained they were one and the same disease. This view seemed convincing to others and many writers based their descriptions and arguments on this supposition.

With time, however, the balance of opinion has turned against such a relationship. Did they follow upon one another, then Riedel's disease would be expected to have an

average later age incidence than Hashimoto's disease. The reverse is the case. Hashimoto's disease is almost entirely confined to females; Riedel's disease has quite an incidence in males, if not as great as in females. Hashimoto's disease is not uncommon; Riedel's disease is rare. It might be argued of course here that only a proportion undergo the transition but the lapse of time since Hashimoto's disease was first described in 1912 has allowed of enough time for observers to note, did the transition occur, the clinical and pathological changes in enough patients to substantiate the thesis. For there is essentially hypothyroidism in the one and a lack of it in the other which is regarded as being the final state. It is almost impossible to imagine that the lack of hypothyroidism in Riedel's disease could follow upon the hypothyroid state which exists in Hashimoto's disease where thyroid cells and vesicles are numerous, although, of course, it is the functional activity of the cell that counts.

Probably many cases are wrongly included under both the Hashimoto and Riedel headings. The most experienced histologists in this work frequently find difficulty in giving definite opinions. In the histological diagnosis of Hashimoto's disease there is the difficulty that aggregations of lymphocytes and lymph follicles are common in all types of goitre, even in hyperplastic toxic glands and in glands of normal size, particularly in older people. These aggregations, although usually more circumscribed in clusters than in Hashimoto's disease, can mimic closely the essentially widespread infiltration of lymphocytes and lymph follicles in Hashimoto's disease. The lymph follicles in Hashimoto's disease tend to show germinal centres and this is regarded by some as peculiar to this disease. But they are to be found also in the lymphoid aggregations of diffuse toxic goitre. The epithelium of the vesicles may show degenerative changes and there is much increase in fibrous tissue.

Levitt (1954) has gone further than other believers in the Hashimoto to Riedel progression and claims that there is an evolution from the epithelial hyperplasia of toxic goitre through successive stages of lympho-epithelial hyperplasia, focal then diffuse lymphoid hyperplasia, fibrolymphoid hyperplasia (Hashimoto) to fibrosis (Riedel's disease). Although all these histological appearances, except for the last, are to be seen frequently, the theory has received little support. Many will deny that they can trace unequivocal evidence of toxicity in the histories of patients with Hashimoto's disease. Too few patients would seem to arrive at the final stage of fibrosis and this stage is certainly reached without the previous clinical phases. That the histologies of Hashimoto's and Riedel's diseases can apparently be seen on the same section in rare instances does not suggest anything more than that Hashimoto's disease may pass in some instances to a degree of fibrosis.

Again, in Riedel's disease, can all cases exhibiting diffuse sclerosis which has not passed beyond the thyroid capsule or at most beyond the adjacent fibres of the surrounding muscle be classed with conviction as Riedel's disease in its earlier stage? Or do they constitute a different entity? The paucity of cases in which the fibrosis infiltrates widely beyond this suggests the possibility of two different ætiologies in the production of the dense fibrous tissue common to both. Certainly the process might stop short from lack of further stimulus at the gland limits, but if it stops short there why should it not also stop short say after involving the whole thickness of the infra-hyoid muscles. It does not seem, from the recorded cases, ever to do so but to go on and envelop carotid sheath and trachea.

The ætiology of Hashimoto's disease is completely obscure. There are no reasons to assume that it is an inflammatory process. Some observers are satisfied that it is preceded by a mild degree of thyrotoxicosis, but it is easy to obtain a collection of symptoms of the "effort syndrome" or "anxiety neurosis" type from women of the age group in which Hashimoto's disease usually occurs. Its usual time of appearance in the later part of the fifth and in the sixth decade with accompanying hyperthyroidism tends more to suggest some exhaustion state or degeneration.

Riedel's disease presents another mystery in regard to causation. While the dense fibrosis suggests a slow chronic inflammation, no help is given from the patients' histories as to any likely antecedent cause or concomitant condition in the way of infection.

Treatment of Hashimoto's Disease, Riedel's Disease and Subacute Pseudotuberculous Thyroiditis. The treatment of these affections is to some extent clouded by the difficulty of accurate diagnosis and, since the differential diagnosis from neoplasm is occasionally difficult, operation is often resorted to unnecessarily if unavoidably.

Hashimoto's disease can, with experience, often be recognized. Since a hypothyroid state is already present an extensive resection will only hasten myxœdema and any resection is therefore uncalled for except in the few patients who exhibit well marked pressure features on the trachea. This form of goitre is particularly radio-sensitive and can be successfully reduced in size by deep X-ray therapy; it is even claimed that the tendency to myxœdema can thus also be halted.

In Riedel's disease, however, pressure features are more prominent and severe and the necessity to free the trachea is more imperative. A resection of the gland, consisting of the isthmus and the anterior parts of both lateral lobes should be done to an extent which will free the trachea. No attempt should be made to dissect out any lateral masses. Laryngotomy and tracheotomy have both been deprecated as palliative methods instead of resection; the relief which they afford, and the difficulty of operation, do not warrant the procedures.

Subacute pseudotuberculous thyroiditis generally arrives at an early spontaneous cure. Where this does not happen then deep X-ray therapy is indicated and is most satisfactory. Thiouracil therapy has also been described as effective; the rationale of this is obscure.

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SIMPLE GOITRE

There can be no positive definition of "simple goitre." The term implies a goitre unaccompanied by toxicity, not malignantly neoplastic, and outside the categories of thyroiditis or Hashimoto's disease. This negative definition is consequent upon the problems of its ætiology which is hardly simple. Yet it is a convenient clinical term to retain for what is by far the commonest type of goitre.

Ætiology and Pathology. Simple goitre may be endemic, occurring in high proportions of the population in certain geographical areas, or it may be of sporadic occurrence in the population of non-endemic areas. The worst endemic regions are Switzerland, the Sierras of Spain and the Himalayas, but nearly all countries possess goitre areas. The most goitrous area in Great Britain comprises the south-western counties.

While there are some discrepancies to be noted in the many investigations of the subject it seems clear that the paramount cause of simple goitre is a deficiency of iodine. In endemic areas (although it is denied for the Himalayas) there is a low content of iodine in the water and in the food grown locally. Only minute quantities of iodine are needed for the normal body economy; the water and soil in endemic areas fail to supply even this much. The explanation of iodine deficiency in mountainous areas is that over the centuries the iodine has been washed out of the soil by rain water free of iodine. This lack of iodine results in a fall in the amount of circulating thyroid hormone in the blood; this in turn stimulates the production of thyrotropin from the pituitary; the increased thyrotropin thereupon stimulates the thyroid and the resulting hyperplasia of the epithelial cells produces a parenchymatous enlargement of the gland. It is probable that in the three main pathological types of simple goitre, parenchymatous, colloid and nodular, there is always a progression from one to the other in that order and that the colloid goitre must be preceded by a parenchymatous one, the nodular by a colloid one. The total amount of iodine in the parenchymatous goitre does not differ significantly from that in the normal-sized gland; presumably the follicles become hyperplastic in order to subtract iodide more efficiently from the plasma.

Although deficiency of iodine, and then usually its deficiency in water and soil, is undoubtedly the chief causative element of simple goitre there must be other factors. The populations of some endemic areas are 100 per cent goitrous and males and females are thus equally affected. In areas of lesser endemicity the percentage is less and females are more affected than males. Why should some of a population drinking the same water and eating the same food become goitrous and others not? Why should the female be more liable than the male? These questions have not been answered satisfactorily. It may be that there is some exogenous factor additional to iodine-deficiency; it may be that there is an endogenous constitutional factor in the individual. Some families in goitre districts go on being goitrous, other families escape.

Additional factors, and some have indeed been put forward as rivals to iodine-deficiency as the chief causative agent, are excesses of calcium and of flourine in the drinking water, as apparently occurs in some but not all endemic areas, vitamin deficiency and intestinal toxæmia Possible endogenous goitrogenic substances have also been thought of in theory; since the disease affects females more often than males an inter-relationship between thyroid and gonads is hypothetically advanced.

Sporadic simple goitre, that occurring in non-endemic areas, may occur at all ages and in both sexes but is much more common in females, especially at puberty, during pregnancy, and at the menopause, all periods when a greater strain is put on the body economy. Those occurring at these times are therefore sometimes called "physiological goitres." Since iodine is ingested in sufficient amount by the population of these areas, it would seem that in some individuals there is a constitutional or endogenous factor which interferes with its synthesis into thyroxin and so determines the appearance of a goitre.

It is often to be noted that in pregnancy a pre-existing simple goitre tends to enlarge. After parturition there is usually only a partial regression is size; with successive pregnancies a step-like rise in size occurs.

Other unusual types of simple goitre remain to be mentioned. Certain vegetables of the *brassica* group, when given to experimental animals in excess or when given without ample other normal foodstuffs, will produce simple goitre. Cabbage is the most familiar of these and the condition is sometimes therefore called "cabbage goitre." It has occurred, although rarely, in humans. The actual chemical nature of the goitrogenic substance in these vegetables and in their seeds is still a matter of argument. Other ingested substances capable of producing goitres, *goitrogens*, are thiouracil, thiourea, sulphonamides, and potassium thiocyanate. The likelihood of the causative element in "cabbage goitre" being of importance in the ætiology of ordinary simple goitre seems to be remote.



(By courtesy of the Editors of the "Practitioner")

FIG. 124. Diffuse parenchymatous non-toxic goitre. Boy aged 9.

The structural changes, which occur in simple goitre are similar in both endemic and sporadic forms. Although there are the three main stages of development with differing typical pictures, it should be realized that any one specimen may show elements of all, the parenchymatous, the colloid and the nodular.

The diffuse parenchymatous enlargement is essentially due to proliferation of the epithelial elements (Fig. 124). The cells lining the vesicles become wider and taller, and may fill up the lumina of the vesicles so that little or no colloid is to be seen. Return to the normal, involution, is possible if iodine be given therapeutically in the early stage of parenchymatous goitre of the endemic variety. Apparent spontaneous subsidence may occur in the sporadic variety. But often in both varieties, whether iodine be given or not, there is a slow gradual transition, probably at different rates in different individuals but in either instance taking a matter of months or years, to the diffuse colloid condition (Fig. 125). The gland remains large or gets larger and the vesicles are now seen under the microscope to be filled out with colloid to beyond normal size; the epithelial cells lining the vesicles are in a single layer and are relatively smaller. To the naked eyes the cut surface of the parenchymatous type appears dark red and "meaty," that of the colloid a golden brown, like honey in the comb. The colloid goitre is relatively poor in iodine. It may be interposed here that the term *colloid goitre* is often used as a clinical description; there is no justification for this, since palpation of a symmetrically enlarged non-toxic goitre cannot reveal whether it is parenchymatous or colloid; the term "non-toxic diffuse goitre" is better. Nevertheless, where a symmetrical non-toxic goitre has been present for several years it is probably in a colloid state. Why this change from a parenchymatous to a colloid state should almost universally occur is not clear. It is easy to understand [illegible]

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intake was never deficient? Possibly the intrinsic constitutional factor referred to above ceases in time to have application and the iodine is again made available to the gland.

The third phase is the nodular state. The diffuse colloid goitre does not seem ever to regress and in most instances develops a lumpiness, usually irregular, throughout the gland. This is due to islands of the gland substance which become isolated and often encapsulated to produce well defined nodules of different sizes (Fig. 127). These usually produce irregular smooth projections on the surface of the gland but occasionally a symmetrically enlarged gland with a smooth regular surface may be found to contain many of them. When they develop in the lower parts of the lobes they tend to grow downwards to produce the most common form of retrosternal goitre (p. 274). These nodules cannot be distinguished, either by the naked eyes or under the microscope, from similar swellings arising in glands which have not apparently gone through parenchymatous and colloid enlargements. These are benign neoplasms, adenomata. The term adenomata is also sometimes applied to the swellings of simple goitre and is objected to by some on the grounds that, arising as they do they cannot be regarded as neoplasms and are better then called nodules. The matter is further considered on p. 268.


Fig. 127. Nodular non-toxic goitre. Female 46.

In a typical nodular goitre there are several, sometimes many, nodules, which may vary greatly in size, some of them reaching great dimensions, as seen in Figs. 127 and 132. Some of them are spherical but some, especially the smaller ones, are oval because of pressure by their more actively growing neighbours. Around the nodules the neighbouring thyroid tissue appears greatly contracted by the pressure of the enlarging nodules; this compressed tissue looks like capsules around the nodules. Through the microscope it is seen that far the commonest nodule is a colloidal one, composed of vesicles (follicles) that in any one nodule usually vary greatly in size, so that the suggested subdivision of the nodules into those composed of large follicles (macrofollicular) and those of small (microfollicular) cannot be supported. Very often in a nodule composed of wide follicles, some of them even cysts, there are areas of very small, closely packed follicles, some of them so small that lumina are not obvious. The majority of the follicles are full of brightly eosinophilous colloid. The cells are usually small or medium sized. While such are the commonest nodules, other types of nodule are occasionally found side by side with them. One that may be called a parenchymatous nodule has no follicles, or only a few, and is composed of solid groups of cells; the solid groups are sometimes in the form of branching cords about three cells thick, the cords being separated by very thin blood-sinuses, the solid cord type of parenchymatous nodule. Some nodules, or parts of the common colloidal nodules, may have a structure approaching very closely that of a



FIG 125. Diffuse colloid non-toxic goitre Female 16 Goitre noticed for 3–4 years. No other symptoms

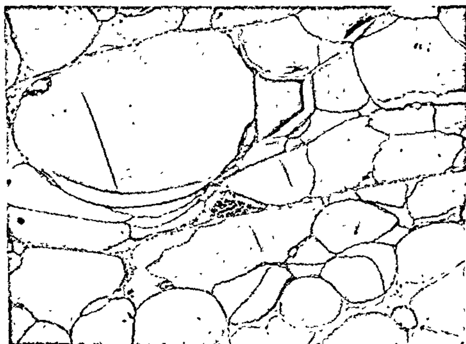


FIG. 126. Diffuse colloid non-toxic goitre Microscopic section showing large vesicles filled with colloid (× 18)

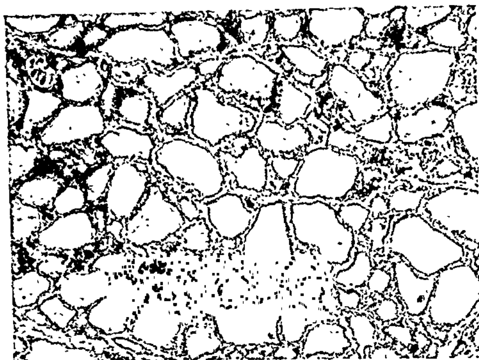


FIG. 128. Thyroid gland Microscopic section of normal thyroid gland (×52).

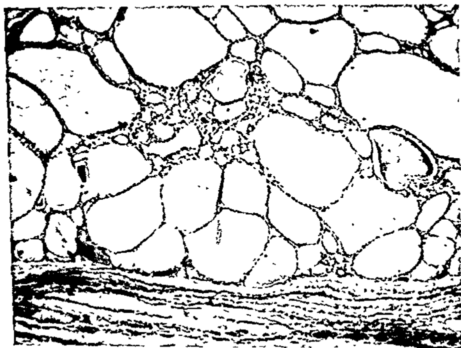


FIG 129. Follicular adenoma (colloid adenoma, male 48). Section through edge of large adenoma showing capsule of compressed fibrotic thyroid tissue (×64).

fœtal adenoma, which consists of very small acini of small cells, some of the acini being close together, others being widely separated by connective tissue which has an appearance of extreme œdema or sometimes of being flooded with colloid. Papilliferous areas may be found composed of wide, even cystic, tubules, with branching intra-cystic papillæ, the tubules being lined, and the papillæ covered, with tall columnar cells. A rather rare occurrence is a nodule composed partly of columnar celled tubules without papillæ and without colloid, and partly of solid groups of large polygonal cells. Occasionally in all sorts of toxic and non-toxic, diffuse and nodular goitres a peculiar cell is seen scattered in ill-defined groups mixed with the other cells; it is conventionally known as the Hürthle cell, though it is doubtful whether it is the cell that was described by Hürthle; its salient characteristic is that compared with the size of its nucleus it has a very large amount of granular cytoplasm that usually takes eosin brightly. Thus, while the colloidal nodule is the commonest, there may be nodules or areas of very different types of tissue in nodular goitres. Hæmorrhage, necrosis, cysts, cholesterol deposit, calcification and severe fibrosis are common in the nodules. Some cysts are retentive, arising from the fusion of adjacent vesicles which are tense to bursting point with colloid; others are degenerative, being due to necrosis or hæmorrhage in a nodule. Masses of cholesterol crystals arise as disintegration products of necrosed tissue. Calcification is an example of the general rule for all tissues, that necrosed tissue is apt to be the site of deposit of lime salts, and wherever there is calcification, bone may be formed. The fibrosis is probably a reaction to degeneration and is seen as irregularly shaped areas of tough white tissue, often hyaline fibrous like fibrocartilage. The degenerations and necrosis are due to failure of the blood circulation in the nodules.

Nodules and Adenomata: Benign Neoplasms of the Thyroid. It is stated above that the swellings or nodules which arise in a colloid goitre to form a nodular goitre are indistinguishable from those which appear to arise as simple neoplasms in a gland which has not, or has not apparently, had any preceding changes. Some pathologists object to the term "adenomatous goitre" being applied to the former, saying that the tumours are not true neoplasms, as apparently they are in the latter. The clinical features of simple nodular goitre and of what they would call true adenomatous goitre are similar and it is convenient therefore to consider benign neoplasms here. But it should first be pointed out that there is still another way in which the tumours may possibly form. That is by hyperinvolution of an area or areas of a gland which has had a short period of overactivity, due to some temporary stress laid upon it. While the affected part or parts of the gland usually involute to normal after this phase, that is to a state of moderate colloid content, an area or areas go beyond this to contain an excess of colloid.

Thus one expects, and finds, that the tumours in the nodular goitre which follows upon a diffuse colloid goitre are multiple and that those which occur by hyperinvolution or are simple neoplasms may be single or multiple.

The *true adenomata* are classified as:

(1) Follicular (colloidal).
(2) Papilliferous.
(3) Solid (parenchymatous).
(4) Fœtal.
(5) Hurthle-celled.

Many pathologists hold that it is impossible with microscopic sections alone to distinguish with certainty between a nodule of nodular goitre whether derived from a diffuse colloid goitre or by hyperinvolution and a true adenoma. Those who claim to be able to do so say that the true adenoma tends to be single, to be better encapsulated, to have the same structure in every part, to be different in structure from the rest of the thyroid and to cause compression of the adjacent thyroid tissue. It is doubtful whether the two lesions are distinguishable on these criteria.

The contention, that if there is only one nodule it is an adenoma, but if there are two then the lesion is a nodular goitre, seems untenable. The thyroid tissue immediately around the nodules of a nodular goitre is certainly compressed to produce good encapsulation. The point about lack of uniformity of structure is inaccurate since the occasional solid parenchymatous nodule in a nodular goitre has exactly the same structure throughout its extent. Perhaps the best criterion would be that in a nodular goitre the thyroid tissue apart from the nodules is abnormal. But this would not necessarily be shown in those described as formed by hyperinvolution and in any case is difficult to assess; the tissue of a diffuse colloidal goitre sometimes does not show in microscopical sections any conspicuous difference from normal thyroid.



Fig. 132. [illegible] age 14. Since then gradual increase in size of goitre No obstructive features. Right vocal cord paralysed. At operation goitre found to be entirely confined to left lobe. Weight of goitre 1,080 grammes.

Radioactive iodine studies have been made to determine the functional activity of the various elements in simple goitre. The radioactive iodine is given and slices of the removed gland are laid against a photographic emulsion. The parts of the gland which have taken up the iodine are thus demonstrated by an *auto-radiogram.* The iodine pick-up in a normal gland is widely distributed. In simple goitre there is a more patchy pick-up, in either parenchyma or nodules, and this seems to be related to those areas in which the follicles are small in size (Fig 131).

Clinical Features. Simple goitre by its name naturally excludes any toxic element. It may, however, be associated with hypothyroidism. Cretins born in areas of high endemicity may present goitres; these result from gross iodine deficiency through several generations and are associated with idiocy and deaf-mutism. Cretins born in non-endemic areas usually have an atrophied thyroid; a goitre may occasionally be present. In areas of less severe endemicity the hypothyroidism is shown by mental and physical backwardness, lethargy, constipation and a tendency to sterility and abortion in women. Again, thiocyanates, thiourea and thiouracil may produce goitres in association with hypothyroidism, but only rarely is simple goitre in non-endemic areas associated with hypothyroid features; such symptoms as may be present are mainly due to pressure of the goitre on adjacent structures, particularly the trachea.

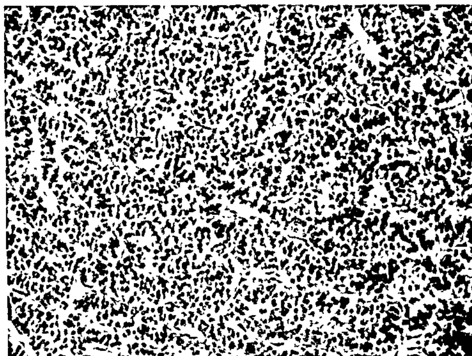


FIG 130 Parenchymatous adenoma. (Solid "cord" type.) Female 41 (×160)



FIG 131 Radioactive study of a nodular non-toxic goitre. Dark areas denote radioactive iodine uptake (×94)

hypothyroidism associated with simple goitre the use of thyroid extract is indicated. Additionally thyroid extract would seem to be beneficial when given to pregnant mothers who are possessed of a goitre. There are rare instances of nodular goitre occurring in childhood in which the giving of thyroid extract seems, by taking the load off the gland proper, to be beneficial in preventing the appearance of further nodules. Radiation treatment, either by the use of radioactive iodine or by external irradiation, has not proved of advantage.

SURGICAL TREATMENT

Surgical interventions in simple goitre are chiefly indicated for the relief of pressure effects and for cosmetic reasons. The first may be a necessity; the second, provided a hardly noticeable scar is inflicted, is reasonable. Resection of the goitre as a prophylactic procedure against the later supervention of toxicity or malignancy is a more debatable procedure. The former complication can be dealt with satisfactorily nowadays; the latter certainly imposes itself more frequently on a nodular gland than on a normal one, but none the less is a rare happening. The subject is dealt with in more detail (p. 308) in the section on malignant goitre. What it is more important to state here is that where there is a single hard nodule in a gland, it may be either a nodule in simple goitre or it may, even if it has been present some time, be a carcinoma. There is thus an indication for the removal of single nodules, based more on the difficulty of diagnosis than on the likelihood of a change of pathology.

Surgical resection is rarely to be recommended for cosmetic reasons before puberty or for a few years after this since the likely sequel is that the goitre will regrow. Should resection, however, be performed thyroid extract should be given until puberty is well past so that the portions left will not have to hypertrophy to produce enough thyroid secretion.

Certain principles are to be observed in partial thyroidectomy performed for simple goitre. Both lobes of the gland must be fully exposed and fully examined. Each lobe should be palpated for nodules. There should be reasonable conservation of the blood supply from the main vessels to obviate the risk of hypothyroid phenomena. The trachea should be completely freed from pressure and be in the midline at the end of the operation. Single nodules should be removed with a good margin of normal tissue lest they prove histologically malignant. Even where there are no clinical or radiological evidences of retrosternal goitre a finger should none the less methodically probe down either side of the trachea for outlying and ectopic masses. Where the trachea is being compressed from both sides the resected portions must include the isthmus of the gland so that the anterior aspect of the trachea is completely bared and free.

The amount of tissue to be resected in operations for simple goitre cannot be laid down; each case must be decided on its merits. Where the operation is being performed for a colloid goitre which is unsightly, an amount equal on both sides should be removed so as to ensure restoration to a neck of normal size and shape; to this should be added removal of that portion of the gland which lies in front of the trachea, whether it be in the nature of an isthmus (which is rarely seen in colloid enlargements) or simply the confluence of the two lateral lobes. Any question of pressure, present or future, will thus be dealt with. All the main vessels should be conserved. A course of thyroid extract therapy, cautiously diminished after a time, will help to prevent regrowth of the goitre

The goitre itself in its parenchymatous form is usually of only small or moderate size. It is soft, symmetrical or roughly so, and rarely exerts pressure on adjacent structures; such pressure is most marked on the infrequent occasions in which the gland encircles the trachea and œsophagus like a collar. Colloid goitres, on the other hand, also symmetrical or roughly so, may be of great size and may cause marked narrowing of the trachea from side to side, the "scabbard" trachea. Their lower poles occasionally come *within the range of being called retrosternal.*

Nodular goitres exhibit variations in clinical appearance from a tiny single nodule which can be seen or felt moving up the neck on deglutition to great masses of nodules projecting from the neck, even overhanging the chest, or on radiological films obscuring the superior mediastinum. The nodules may be present in only one lobe when, if of any size, they will displace the trachea and larynx to the opposite side, causing a variable degree of narrowing in the former. This may be as severe as to cause dyspnœa; a patient with much tracheal displacement who falls asleep on the side to which deflection is occurring may assume a position which accentuates the narrowing and wake suddenly struggling for breath. Other pressure effects are uncommon where the swelling is confined to the neck and does not extend retrosternally, the soft superficial tissues of the neck allowing of outward expansion. Pressure on pharynx and œsophagus is as uncommon under these circumstances as to make one suspect any complaint of dysphagia as being functional in origin, for allegations that the food "seems to stick" are as common with small as with large cervical goitres, although uncommon in both instances. The greatest pressure effects occur in retrosternal goitres (p. 274).

Treatment of Simple Goitre

Prophylaxis

The obvious possibilities of iodine therapy as a prophylactic agent against goitre formation has led to large-scale experiments in endemic areas. While the results have not been uniform, such great successes have been achieved that even in the United Kingdom, a country with areas of low or only moderate endemicity, prophylaxis is to be tried on a national scale by the introduction of a small quantity of iodine into all table salt. The use of iodine therapy after a goitre has appeared is of little value; the purpose must be to prevent the occurrence of parenchymatous goitre, the first stage in simple goitre.

In endemic areas therefore, it can well be understood, the need for iodine is especially high in pregnant women, so that their offspring will be unaffected, and in children and adolescents, to meet the demands of the period of growth. While several methods of administering iodine have been tried, more success has been attained by the use of iodised table salt than by the others since the systematic taking of any medicines is apt to go by default, whereas salt, although some children are apt to avoid it, is commonly taken at all meals.

Medical Treatment

While the ingestion of iodine may occasionally seem to cause recession and disappearance of a small goitre in the parenchymatous stage it has in the main little or no effect on an established simple goitre. Indeed, although it is now less asserted, prolonged iodine therapy is regarded by some as tending to initiate thyrotoxicosis. Where there is obvious

Other retrosternal goitres may occasionally develop from nodule formation in thyroid tissue which has little or no apparent connection with the main gland, which itself is almost invariably nodularly enlarged. Such ectopic portions of tissue, *mediastinal aberrant goitres*, may exist down either side of the trachea; their blood supply comes from above.



FIG. 133. X-ray showing narrowing of trachea from side to side caused by a nodular non-toxic goitre

Very rarely the whole gland may be intrathoracic and swellings occurring in such a gland will present no swelling in the neck even on deglutition.

Clinical Features. Because of the unyielding bony framework of the upper chest the pressure effects are much more pronounced than are those of a goitre confined to the neck. The deviations and narrowings, lateral or antero-posterior, of the trachea may be pronounced. Shortness of breath and dyspnœa, even stridor, may result. Suffocative attacks during sleep are common and an irritative cough may develop. It is remarkable how rarely dysphagia occurs despite obvious pressure upon, and deviation of, the

and delay nodular formations in the remaining portions. Additionally it will prevent the possible appearance of hypothyroid phenomena, in particular the acquisition of too much weight.

In nodular goitre the problem is much more varied and may be difficult. It is easy where there are a few discrete nodules or a circumscribed nodular mass; each is removed by resection-enucleation, a term which implies removal of the nodule or mass with a thin slice of adjacent thyroid tissue. Where, however, there are many nodules in both lobes, and some glands may be riddled with nodules, often in all stages of degeneration, then it seems impossible, and often is impossible, to clear the gland of all its nodules and at the same time leave much normal thyroid tissue. Indeed there are nodular glands which, if all nodules are to be eliminated, would require total thyroidectomy, and such has even been advised. There is, moreover, the question of recurrence, the development of nodules from seedlings in the remnants. A careful survey of the whole gland, before any resection is started, is therefore advisable, particularly to determine where there is any reasonably sized areas of normal tissue which can be conserved. It is hardly sufficient to take the attitude that conservation matters little since thyroid extract can be easily given to make up any deficiency; the taking of the extract can be irksome to patients, the amount required takes adjustment, and some supervision has to be maintained over patients who are taking it. It will quite often be found that the only normal tissue in a highly nodular gland is in the region of the superior pole, and that it is possible to conserve this portion with the superior thyroid vessel intact.

Where both lobes are riddled throughout with nodules most surgeons adopt a compromise, retaining moderate stumps of admittedly doubtful tissue, doubtful both as to function and as to whether they contain seedlings which may grow to produce recurrences.

The tying of main vessels, a feature of importance in the operation in toxic goitre, should not be lightly done in simple goitre. It may, however, be necessary at times to divide the superior vessels in order to mobilize a lobe where the superior part has necessarily to be resected. It may also occasionally be convenient to ligature the inferior artery where the whole of the lower part is to be resected. Ligature of vessels, on the argument that diminished blood supply will prevent the formation of further nodules, does not commend itself to the writer.

Retrosternal Goitre. The lower poles of the lobes of uniformly enlarged glands, such as those of colloid goitre, will project on occasion into the thoracic inlet and thus be partly intrathoracic, although moving freely up and down in the inlet. The term *retrosternal goitre*, however, is mainly reserved for those glands in which the main mass of the goitre is intrathoracic, or, more commonly, where, although there is a cervical mass, there is an intrathoracic portion connected to it by a neck and, by its bulk, incapable of escaping from the thorax on deglutition, as happens also where the main mass is intrathoracic. (The terms *substernal goitre* and *intrathoracic goitre*, sometimes used, are essentially synonymous with *retrosternal goitre*.) An uncommon intermediate type, *goitre plongeant*, signifies a small more defined intrathoracic goitre which is forced jerkily up into the neck on deglutition, to recede into the chest almost as quickly when the expulsive force is spent.

Occasionally an enlarging cervical goitre breaks through between the infrahyoid muscles and enlarges downwards in front of the sternum instead of passing into the retrosternal space (Fig. 132) in front or behind the trachea, more commonly the former.

œsophagus as seen in X-ray films when barium is swallowed. More striking are the effects of pressure on the large veins, in particular the two innominates and the superior vena cava. This embarrasses the venous return from the head, neck and upper extremities. As a result the superficial veins of the neck and of the upper chest wall enlarge in order to carry the blood back to the heart by way of the intercostal and azygos veins. In the most pronounced instances of this there is cyanosis and even œdema of the head and neck and upper extremities. The recurrent laryngeal nerve is sometimes affected; rare instances have been reported of pressure effects on the sympathetic and phrenic nerves.

The *mediastinal aberrant goitres* occupy the same positions as the retrosternal prolongations of cervical goitres and cause the same pressure features. If at all goitre operations a routine practice be made of passing a finger down through the thoracic inlet along each side of the trachea, such masses will be found more frequently than symptoms or radiology suggest.

A nodular mass may be removed from one lobe in the neck while a retrosternal prolongation of the other lobe is the cause of the symptoms.

The retrosternal extensions generally pass into the anterior part of the superior mediastinum, only rarely into the posterior. Respectively they are sometimes loosely called anterior and posterior mediastinal goitres; if they project far enough downwards they will enter the anatomical anterior and posterior mediastinal spaces, but before they reach these they are really in the superior mediastinum. The anterior passes in front of the trachea, the great vessels and the recurrent nerve. The rare posterior one, more frequently found on the right than on the left, lies behind the lines of these structures, and may have the carotid sheath and inferior thyroid artery in front of it; the trachea and œsophagus are usually displaced laterally but the former may even be displaced anteriorly. The different courses of these retrosternal extensions can be explained by the site of origin of the mass in the thyroid itself.

Retrosternal goitres are rarely associated with toxicity (Fig. 136).

The diagnosis of retrosternal goitre, while it may be made frequently with confidence on clinical grounds, requires confirmation and elaboration by radiology, when, in particular, the deformations of the trachea can be studied. Because of a complaint of pressure symptoms the thoracic inlet is often X-rayed although little or no goitre is seen or felt in the neck. An opaque mass in the retrosternal space may be seen and the question is raised as to whether it is goitrous, aneurysmal, or neoplastic. If no goitre can be felt in the neck and no swelling seems to come up through the inlet on deglutition then the mass seen on X-ray is unlikely to be a retrosternal goitre; it is more likely to be an aneurysm of the aorta or a malignant thoracic neoplasm. Thymic tumours present a characteristic rectilinear outline on X-ray and do not cause the same amount of difficulty of interpretation.

Treatment

Retrosternal goitres should always be removed. Even if symptoms are slight the likelihood is that they will increase since the mass usually enlarges. Again, the nodular masses are often cystic or soft and hæmorrhages into them are common. Asphyxial attacks have occurred from such incidents, and the possibility of such a cause of asphyxia should be borne in mind when the catastrophe occurs in someone not known to possess a goitre.



Fig. 134 Retrosternal goitre. Male 58. Goitre present for 27 years. At operation the left lobe of the thyroid was larger than the right and almost entirely retrosteral.



Fig. 135 Nodular retrosternal goitre

Their removal, a procedure that often looks formidable, is not usually a difficult matter. Except for the very rare instances of totally intrathoracic goitres and so-called posterior mediastinal goitres, the operation can be performed through the usual neck exposure without any splitting of the sternum or other opening of the chest wall. It is to be noted that the retrosternal mass is usually nodular and relatively avascular and that



FIG 138 X-ray of thoracic inlet showing displacement and narrowing of trachea by a large retrosternal goitre Male aged 61. Goitre noticed for 25 years.

its blood vessels descend with it, so that, if necessary, the inferior arteries may be tied before bringing up the mass. After exposing the anterior aspect of the gland in the neck the operator works laterally under the infrahyoid muscles to reach the sulcus between gland and carotid sheath. If a finger be now passed downwards into the chest in this artificially made groove the mass can usually be prised gently out of the retrosternal space. Where the goitre in the neck is large and the whole mass is jammed, then division of the superior vessels, mobilization of the upper pole and a certain amount of freeing

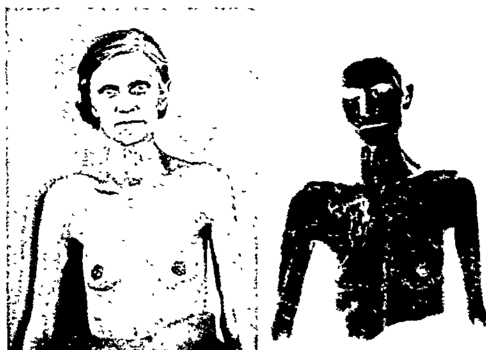


Fig. 136. Retrosternal secondary toxic goitre. Female aged 47. Goitre for 20 years. Toxic symptoms present for 8 months Right lobe chiefly enlarged and partly retrosternal. Right vocal cord paralysed Right phrenic nerve also paralysed. The infra-red photograph shows the distended veins in the right chest wall.



Fig. 137 Chest X-ray of same patient as in Fig 136. Note the large retrosternal goitre. The right dome of the diaphragm was paralysed.

obtained. Where a history of mental stress is elicited it is remarkable that the emotion or pity aroused in the patient is almost always in respect of someone else.

The rare hyperfunctioning adenoma (p. 285) suggests that in some instances the stimulus to overaction comes from within the gland itself.

The sex incidence of toxic goitre is approximately five females to every male. The disease may occur at any age, but is most common in the fourth and fifth decades. It occurs in all races.



FIG 139 Diffuse toxic goitre Female age 16 Symmetrically enlarged gland Marked exophthalmos and lid retraction.



FIG 140 Nodular toxic goitre Female age 68. Notice the marked wasting and absence of eye changes

Classification. Much confusion is caused by the many varied terminologies used. A simple clinical classification, for reasons to be elaborated, is:

1. *Diffuse toxic goitre* (inclusive of, or synonymous with *primary toxic goitre, exophthalmic goitre, Graves' disease*).

2. *Nodular toxic goitre* (inclusive of, or synonymous with *secondary toxic goitre, toxic adenomatous goitre, toxic adenoma*)

Many criticisms can be made of any classifications. What is implied by *diffuse toxic goitre* (Fig. 139) is the association of thyrotoxicosis with a thyroid gland which is smoothly and uniformly enlarged and which gives an impression on palpation that if it were cut into it would be homogeneous throughout. "Exophthalmic" goitre and Graves' disease imply the presence of abnormal ocular physical signs which are not always present and are not essential to the diagnosis. What, on the other hand, is implied by *toxic nodular goitre* (Fig. 140) is the combination of toxic features and a thyroid gland which has a palpable nodule or nodules. It cannot be said from clinical examination, it can often not even be determined from the portion of gland removed at operation, whether the toxicity

from the trachea will allow of the whole mass being pulled upwards. A further manœuvre, admittedly somewhat messy, may be required where the mass has expanded considerably below the thoracic inlet. Since the mass usually consists of mushy nodules or cysts, a finger pushed into its centre can produce a flow of fluid or soft jelly-like material. This effects such a diminution in bulk of the mass as to allow a finger to be passed around the outer aspect of its capsule and thus to lever it up and out. The inferior thyroid artery, the wall of which is often degenerate from long continued pressure, should be tied in continuity at some point in the operation.

When, however, it is recognized at operation that a large mass is passing down in the posterior part of the superior mediastinum, then the dangers to the recurrent nerve and arteries may necessitate splitting of the manubrium sterni down to, and with a crosscut into, the second rib interspace. This will increase the diameter of the thoracic inlet and allow the mass to be pulled gently upwards with less likelihood of damage. The inferior thyroid artery should be sought for early and divided between ligatures.

The rare totally intrathoracic goitres have to be dealt with by thoracotomy.

The space left after the removal of a large retrosternal goitre takes time to be obliterated by the expansion of the upper parts of the lungs. Drainage is required for several days. The space fills with clot and X-rays will show that this does not clear for three or four weeks.

TOXIC GOITRE

(Thyrotoxicosis)

Toxic goitre is a comprehensive term embracing several varieties of a disease in which, due to an excessive secretion of thyroid hormone, there are widespread over-stimulating effects on the various tissues and systems of the body. These result essentially in an enhanced metabolism; there is, so to speak, a burning-up of the body tissues at a rate which cannot usually be made good. The circulatory and nervous systems are also particularly affected.

Ætiology. The ætiology of thyrotoxicosis is unknown. The most generally accepted theory is that in most instances some abnormal stimulus acts upon the hypothalamus which in turn stimulates the anterior pituitary to produce excess of thyrotropic hormone. This in turn produces an excess of thyroid hormone. There is no evidence that the nature of the hormone is altered. But, since excess of circulating thyroid hormone should inhibit the amount of thyrotropic hormone produced there must in toxic goitre be some breakdown of this regulator mechanism.

The nature of the stimulus or stimuli acting upon the hypothalamus is still uncertain. There would seem to be in many of the patients a failure of adaptation to stresses, physical and mental. Thus the disease seems in many instances to follow upon infections, such as acute tonsillitis or the exanthemata, upon pregnancy and parturition, and upon psychical disturbances, sudden or prolonged. That most people pass through these stresses without developing thyrotoxicosis does not invalidate the argument, if one allows that there are constitutional types prone to certain diseases. Certainly the relationship of the onset of thyrotoxicosis to these alleged stresses is often striking. But it may well be, and careful detailed history-taking will often bring it out, that the disease was present in mild degree before the stress occurred; the stress caused accentuation of already existing features. In many patients no history of physical or mental strain can be

the degree of toxicity. Occasionally one lobe is larger than the other. The colour is commonly a reddish brown. In consistence the gland is usually firm but may be hard and friable, especially after prolonged iodine medication. The cut surface shows a varying picture depending upon the colloid content, the vascularity and the amount of fibrous stroma. Where no iodine has been given the surface is red and meaty; where iodine



FIG. 143 Microscopic section of thyroid gland in untreated diffuse toxic goitre The dark areas are lymphoid tissue (×18)

medication has re-established a colloid content the colour is more of a deep golden. The rare opportunities of examination of a highly hyperactive gland not subjected to iodine therapy show that histologically the follicles, perhaps only in some areas, are hardly identifiable, so great is the proliferation of their lining cells which increase in size and height and become raised on papillary projections which obliterate the lumen of the vesicle (Figs. 143, 144). What distinct follicles are seen are usually smaller than normal. In less hyperactive glands the picture is not so severe. After iodine administration the follicular structure is largely regained by the resumption of colloid storage, and the lining cells decrease in size and height (Fig. 145). Thiouracil administration, on the other hand, tends to preserve or increase the hyperplasia. Aggregations of lymphocytic cells may be seen in sections of glands producing all degrees of toxicity.

is derived from the nodules or from the rest of the gland. Some nodules in a removed specimen show such degenerative changes as to rule themselves out as a source of toxicity; nodules in another specimen may look more active than the rest of the gland and suggest themselves as the source. Radioactive iodine studies have confirmed that the over-secreting cells may in some instances be in the nodules, in others in the gland proper. The term *toxic adenoma*, not uncommonly used, should therefore be discarded from clinical use as should the term *secondary toxic goitre* which would attribute causation to the presence of the nodules.



FIG 141 Diffuse toxic goitre Female age 50 Showing disparity in size of lobes and lid retraction only



FIG. 142. Nodular toxic goitre with symmetrically enlarged gland Female age 57

Mistakes may frequently be made in the clinical separation between the two types set out In diffuse toxic goitre, for instance, one lobe may be much larger and more palpable than the other (Fig 141), this gives rise to the erroneous diagnosis of toxic nodular goitre. Again the gland may be symmetrically enlarged and smooth of surface, yet on section each lobe may contain many nodules (Fig 142)

Whether or not the two types are distinct diseases or whether they constitute the same disease but with the stimulus falling in the one instance on a normal gland, in the other on an already nodular gland, is a matter of argument, and some possible differences will be noted later. But it is evident that a clear distinction cannot always be made with certainty on clinical examination although a strong presumptive one may be offered. It is convenient, none the less, to retain the classification.

Pathology

In *diffuse toxic goitre* the gland is usually uniformly enlarged and retains the same general shape. The enlargement may be slight or great but size bears no relationship to

In *toxic nodular goitre* the naked eye appearances usually resemble those of the non-toxic nodular form, but where the nodules are few the mass of the gland may resemble that of the toxic diffuse form. The nodules may present all the variations described in the non-toxic form. In some instances it will be clear to the eye whether the over-production of the hormone has been taking place in the nodules or in the gland tissue proper but in most this will not be obvious. There is a rare type, the hyperfunctioning adenoma, in which a single nodule, or it may be two, not only over-secretes to produce toxicity but takes over the function of the gland proper so that the latter becomes dormant in activity, showing this by pallor and shrinking. Removal of the hyperfunctioning adenoma produces at first marked hypo-thyroidism; gradually the gland resumes its function and equilibrium is restored.

General Picture and Course of the Disease

Since the excess of thyroid hormone reaches all the cells of the body it is not surprising that a wide range of symptoms and signs may be produced. Nevertheless the picture varies from patient to patient, the process sometimes quite selective as to which systems are chiefly affected. Thus loss of weight will be the predominant complaint and obvious sign in one patient, other features being hardly discernible. In another, eye changes, tremor and obvious excessive perspiration will present, with little or no loss of weight.

Because of the increased metabolism a salient characteristic is loss of weight despite an increased appetite. That there are exceptions to this does not invalidate the combination as the most significant item in the history.

Before individual features are discussed it may be said that broadly there are two main clinical pictures of the disease, the differences between which tend to be seen at their clearest in the young and in the elderly. In the young, who in the main will naturally have glands which are diffusely enlarged, patients suffering severely or moderately severely present marked nervousness, tremulousness, excessive perspiration, exophthalmos and tachycardia. In the elderly sufferer the goitre is commonly nodular and the features tend to be more those of lassitude, loss of weight and a tendency to heart irregularities; eye changes are often absent and, if present, lid retraction is more common than exophthalmos. While these two broad clinical types are to be noted there are so many exceptions in regard to age and variation as to symptoms that they must not be over-emphasized.

The disease has usually been developing slowly for several months before advice is sought. The patient is not unduly troubled for a time by symptoms which in his or her mind can be attributed often to other causes. Not a few, especially younger patients, feel at first even fitter and more active and it is only on the supervention of eye features and the development of an obvious neck swelling that they, or their friends, notice something amiss On the other hand the patient may present with auricular fibrillation and little else.

If untreated, the disease tends usually to persist for a period of years, waxing and waning in severity of symptoms. The final issues, if the disease be untreated, vary. The disease may spontaneously disappear. Physicians of the past treated their patients with rest and sedatives and claimed that in time the disease often extinguished itself. While no doubt this did happen occasionally, it is doubtful if the event was as frequent as they thought and there are no long-term follow-up results to confirm their opinion. It is



FIG 144 Lymph follicle with "germ centre" in diffuse toxic goitre (Graves' disease) Female age 30 (× 180).



FIG 145 Diffuse toxic goitre after iodine therapy Note the follicles and colloid storage. Male age 25 (× 52)

There is additionally a volatility about the pulse rate that is not present in normal persons except the most nervous. While the rate during sleep is often less than the habitual rate, the discrepancy is not as marked as in nervous tachycardia. Slight incidents, exertions and irritations make the pulse rate soar and decline in bizarre fashion. Put to rest in bed, the average thyrotoxic patient after a day or two usually experiences a drop in the pulse rate of ten to twenty beats per minute, the new rate however still remaining significantly above normal. A moderate rise in the systolic blood pressure is a feature of thyrotoxicity, which may also in some instances be associated with a hypertension due to the toxæmia. Ordinarily the pulse pressure is raised.

Radiology in thyrotoxicosis shows a moderate enlargement of the heart. An electrocardiogram may show arrhythmics and other changes.

The *nervous and mental features* to be elicited in the history are, typically, increasing nervousness and irritability. The nervousness frequently results in an unwillingness to meet and move among other people; the irritability would seem often to be due mostly to the patient's consciousness of not being as fit, competent and robust as she was. Tremulousness is a common nervous feature, shown often by the dropping of crockery or the upsetting of their contents, or in irregular handwriting. The condition is only partly nervous; muscular weakness is also a component. Objectively, a fine tremor of the outstretched hands and fingers is said to be the characteristic type, but often where the disease is severe the tremor is gross and coarse. Choreiform movement of the hands and arms usually signify a severe degree of the disease; they are seen particularly well in children. But, short of these, fidgety movements are highly characteristic of thyrotoxic patients, particularly younger ones. In this connection it may be noted that, apart from those with decompensated heart disease, patients tend to display a combination of intermittent energy and fatigue.

Psychoses may occur. There is no psychosis typical of the disease and, presumably depending on the mental make-up of the patient, it may be manic, schizophrenic or paranoid in type, to mention the three more common. A psychosis usually indicates a severe degree of intoxication.

One of the effects on the *digestive system*, increased appetite, has already been mentioned. Another effect is a tendency to looseness of the bowels amounting in severe cases to copious diarrhœa. However, women normally tend to be constipated and the only effect may be that a more regular action is achieved; such a point in history-taking is of value. Nausea and vomiting, when they occur, are signs of serious intoxication. An associated glycosuria, which is neither diabetes mellitus nor renal glycosuria, is occasionally found, and is of no great significance; it is due to an increased tolerance for glucose.

Eye Features. While eye signs are common, their absence does not rule out the diagnosis. Again, exophthalmos may be due to other causes and there is a particular form in which may be associated with a goitre producing little or no toxicity; in such cases the exophthalmos may be very pronounced and be accompanied by paresis of some of the ocular muscles. Such a condition (p. 305) is variously described as thyrotropic exophthalmos, malignant exophthalmos, and exophthalmic ophthalmoplegia.

The [illegible] 46–147). The [illegible] present. Exophthalmos entails a pushing forward of the eyeball, a proptosis. In lid retraction, which is thought to be due to a spastic state of the levator palpebræ superioris, the upper

more likely that in many instances the disease still smouldered, maybe to re-erupt in severe form, maybe eventually to produce auricular fibrillation, the result of a low grade thyro-intoxication of the heart muscle over many years. The untreated patients with thyrotoxic auricular fibrillation eventually pass into heart failure.

Sufferers from the more acute form of the disease may, whether treated or untreated, undergo a toxic crisis, a "thyroid storm," in which all nervous and cardiovascular features are grossly accentuated and in which state the patient is likely to die.

Symptoms and Signs in Detail

In many sufferers the diagnosis is easily determined, especially if eye changes are present. But many patients possessed of a goitre complain of symptoms which they and their doctors are too easily inclined to attribute to the swelling in the neck. Additionally, patients may complain of symptoms highly suggestive of thyrotoxicosis and yet a goitre may not be seen or sufficiently well felt to warrant the diagnosis. This discussion of symptoms and signs is therefore made to underline the most significant points in correlation and, above all, to distinguish real from functional symptoms. Even eye changes may be misleading.

While weight loss as against an enhanced appetite is most significant in the history, such weight loss is not usually obvious, most people being able to lose one or two stones without showing it. On the other hand, gross emaciation may occur (Fig. 140). Young patients may actually increase in weight, their intake due to their ravenous appetite exceeding their loss from increased metabolism. Appetite is an individual and capricious thing and some will not admit to its exaggeration; where a good appetite is admitted, however, it is significant. While complaint of a poor appetite tends to invalidate the diagnosis there are again exceptions; elderly patients particularly may disclaim a good appetite; the presence of heart failure may depress the appetite as may also the digitalis given to counteract the failure.

The enhanced metabolism further shows in increased perspiration, in a feeling of warmth, in a preference for cool weather. The thyrotoxic tends to keep away from any fire or stove, needs few bedclothes, has soft rather moist warm hands. Only in exceptional cases is there a raised temperature. A preference for cool weather, however, is by no means universal; it would be strange if there were no exceptions to the rule in a matter which is so variable in healthy people.

The effects on the *cardiovascular system* show themselves subjectively mostly in the form of attacks of palpitation, commonly on exertions which previously would not have produced it, even more significantly occurring at rest, and then sometimes interrupted by "missed beats" signifying extra systoles. Cardiac pain is seldom complained of. Subjectively also there may be attacks in which the palpitation is accompanied by a consciousness that the heart is racing, and by sensations of fright, giddiness and uncertainty. During such attacks, which signify episodes of paroxysmal tachycardia or auricular flutter, the patients cannot pursue their ordinary occupations. Additional consciousness of irregularity of the racing pulse signifies paroxysmal auricular fibrillation. But continuous fibrillation may set in without such disturbing episodes and the symptoms of uncompensated heart disease appear gradually.

The patient may or may not be conscious of a rapid heart action, as distinct from the forcible beats of palpitation but tachycardia is an almost constant objective feature.

symmetrical goitres; in toxic nodular goitre they occur only occasionally and then lid retraction is more common than exophthalmos.

Subjectively, pain behind the eyes is a common complaint when exophthalmos is present; watering of the eyes and photophobia are not infrequent. In severe exophthalmos the cornea, owing to the lids failing to cover it, may become ulcerated.

The theories of the causation of exophthalmos are dealt with elsewhere (p. 306).

The *size of the goitre* is unrelated to the degree of toxicity or exophthalmos. What may be found in the examination of goitres in the more severe instances of toxicity are signs of great vascularity as shown in palpable thrills and audible bruits. Their presence signifies great overactivity of the gland. The gland may, particularly in men, prove at operation to be much larger than is thought on clinical examination (p. 292).



Fig 148 Pre-tibial myxœdema. Pre-operative photograph in a female age 29 with a diffuse toxic goitre

Muscular weakness is a common feature and is the cause of the easy exhaustion of which most patients complain. Additionally it is partly the cause of tremor. Exceptionally, and this is almost always in young patients, there is no such complaint, a highly active and even strenuous life being followed although other symptoms and signs of the disease are well marked. Generally, however, this is a matter of determination of a patient not to surrender to any feeling of weakness. The muscles participate in the general wasting.

Sometimes a muscle or group of muscles wastes out of proportion to the rest of the muscular system. *Chronic thyrotoxic myopathy* affects most commonly the muscles of the shoulder and pelvic girdles. In *acute thyrotoxic myopathy*, a rare condition, the bulbar muscles, among others, may be affected. Rare cases of periodic paralysis may be associated with thyrotoxicity.

Creatinuria occurs in thyrotoxic patients and a defect in the metabolism of creatine is held to be the cause of the weakness and wasting since creatine is required for the contractions of skeletal muscle. It has been found that the administration of iodine or thiouracil, or the performance of thyroidectomy, abolishes creatinuria in toxic goitre, whereas the creatinuria of myopathies unassociated with thyrotoxicity is unaffected by these measures. The myasthenia of toxic goitre may simulate myasthenia gravis but there is a lack of response, although not without exception, of the former to prostigmine. The myopathy of exophthalmic ophthalmoplegia is referred to on p. 305.

Osteoporosis of bones has been noted from time to time in thyrotoxicosis and its cause is disputed. One view is that the thyroid hormone acts directly upon the bones, releasing their calcium and phosphorus; another is that where osteoporosis occurs there is an associated hyperparathyroidism; still another is that the renal threshold for calcium is lowered as an effect of the excess of circulating thyroid hormone on the kidney.

eyelid is kept elevated but there is not necessarily any associated proptosis. A rough test depends upon the fact that the upper and lower eyelids usually, but not invariably, impinge on the upper and lower parts of the iris in normal people. Where lid retraction alone is present, a band of sclera is apparent between the margin of the upper eyelid and the iris but the lower eyelid still impinges on the iris. When proptosis occurs a band of



FIG 146 Diffuse toxic goitre. Female age 41 Note the staring expression



FIG 147. Diffuse toxic goitre with unequal exophthalmos Female age 32

sclera becomes apparent between lower lid and iris and then between upper lid and iris. But exophthalmos and lid retraction often co-exist.

Both these signs are unmistakable when well-marked. Their lesser degrees, however, are to be sought for since their importance in clinching a diagnosis is great. A staring or frightened expression (Fig. 146), especially when such has been confirmed as a change by friends or relatives, if often highly significant. The staring expression seems to be due to a mild degree of lid retraction with infrequent winking, or it may be due to a degree of exophthalmos sufficient only to widen the palpebral fissures slightly. Many tests and signs have been propounded to show or confirm ocular changes. Most are of doubtful value but Von Graefe's sign, a lagging behind of the upper lid as the eyeball is made to travel downwards (they normally move concurrently), confirms the less well marked degrees of lid retraction.

While lid retraction is nearly always of equal degree on the two sides, exophthalmos not infrequently first becomes apparent in one eye and may be quite marked before the other eye becomes affected (Fig. 147) The degree of proptosis usually in time becomes equal on both sides.

Diplopia is not uncommonly associated with exophthalmos, without there being any suspicion of the condition of exophthalmic ophthalmoplegia. Swelling of the eyelids may also accompany exophthalmos as also may chemosis of the conjunctivæ.

Eye features are more common in the young and in those with smooth, that is, diffuse

Radioactive Iodine Uptake Test. This is dependent upon the amount of iodine excreted in the urine after the administration of a standard dose of radioactive iodine to the patient. The more active the gland the more iodine will it take up and correspondingly less will be excreted in the urine. Here again, however, as in B.M.R. estimations, there is a range of variation in normal people and in thyrotoxic patients, and the figures for one overlap the figures for the other. In round figures the urinary excretion of radioactive iodine in these tests varies in the healthy (euthyroid) from 25–95 per cent, with a mean of 60; in the thyrotoxic patients the range is from 10–45 per cent with a mean of 25 per cent.

The usual method is to estimate the amount excreted in the urine after taking a known dose by mouth.

Another method commonly employed is to calculate the rate of uptake in the thyroid gland by placing a Geiger-Müller counter over the neck after administration of a solution of radioactive iodine. The same objections hold as in the radioactive iodine excretion test.

Other methods, which have been employed include comparing the uptake in the neck with the bloodstream and estimating the radioactive protein bound iodine in the blood.

Protein Bound Iodine. Although this estimation gives a direct measure of the level of circulating thyroid hormone in the bloodstream, the technique is very difficult, expensive and requires much experience.

Blood Cholesterol. Generally speaking the level of blood cholesterol falls in hyperthyroidism and rises in hypothyroidism. The estimation is therefore of some value, but as in B.M.R. estimations it is of greatest value in a negative sense in hyperthyroidism; thus a high blood cholesterol figure is more against hyperthyroidism than is a low one in its favour. The variation in the amount of blood cholesterol in healthy beings is a wide one; any figure from 100–300 mg. per 100 ml. may be normal. Accordingly there is, the normal for the individual in health being unknown, too great a range for the figures to be helpful where diagnosis of thyrotoxicity is in doubt.

Differential Diagnosis

From the innumerable features which toxic goitre may present it might be thought that diagnosis need present little difficulty. This is true for the majority of patients but there remains a sizeable minority who present the problem of diagnosis of toxicity in different ways. Where there are definite eye signs the other features usually, though not invariably (see p. 306), fall into line.

The chief diagnostic difficulty lies in the simulation of many of the protean manifestations of the disease by functional states which are variously called anxiety neurosis, effort syndrome, nervous tachycardia, neuro-circulatory asthenia and suchlike. In these there is a combination and presentation of such features as tachycardia, nervousness, fatigue, lassitude, weight variation, and a tendency to excessive perspiration. That a goitre is present does not mean the symptoms emanate from it. Simple goitre is common and it is only too easy to hold it blameworthy. The most significant points of difference would seem to be that the "anxiety neurosis" complains of a poor appetite, that she has cold clammy palms, and that her history over the years gives other evidences of functional disturbances. But functional symptoms often accompany real ones, especially in the milder forms of the disease. While it might be argued that if the patient has a goitre no

"*Pretibial Myxœdema*" (Fig. 148). A coarse localized bilateral leathery thickening of the skin of the fronts of the legs is a rare complication, although it may appear some time after successful thyroidectomy for the disease. The affected area is shiny and pinkish or brown in colour, with a peau d'orange resemblance, and may have coarse hair. Biopsies have shown that it is due to a deposition of mucin between the connective tissue fibres of the cutis. It has no relation to the degree of hyperthyroidism but seems to have some connection with whatever causes exophthalmos. The condition causes little trouble other than cosmetic ugliness. Thyroidectomy and thiouracil therapy have no effect on it. The tendency is towards spontaneous disappearance or lessening.

Laboratory Findings in Thyrotoxicosis

Many laboratory tests are and have been employed in the diagnosis of thyrotoxicosis. Estimations of the basal metabolic rate, of radioactive iodine uptake and of blood cholesterol are those most commonly made. But there are many others. Their purposes have been to add to our knowledge of increased metabolism, to assist in determination of the degree of toxicity, and, where toxicity is doubtful on clinical grounds, to help clear the issue. The drawback to the last of these, which from the surgeon's point of view is the most important, is that healthy people exhibit a range of variation to such laboratory estimations. Since it is the lesser degrees of toxicity which are the most difficult to determine clinically, a variation from an individual patient's normal (which naturally was not estimated during health) may not pass, when toxicity is present, beyond the range of normality for all. On the other hand, however, the doubtfully toxic patient may show an estimation which definitely rules out toxicity, and this is most helpful.

The most that can be said about laboratory tests is that they can, when viewed alongside the clinical findings, be of value. But the diagnosis must not be made on any one of them or on a combination of them alone.

The *basal metabolic rate* (B.M.R.) is raised. While the "normal" figure for an individual is taken as 100 per cent no abnormal significance can be attached to readings between 85–115 per cent since these may be found in healthy individuals. It is not unfair to point out that the readings made in many institutions are unreliable. Unless the estimation is made by a competent technician of great experience, working under the best conditions, the figure will frequently be erroneous. Errors are almost invariably on the side of over-estimation. Where a department has made a special study of the subject and makes repeated readings on the same patient, then the assessment may be highly accurate. An isolated single reading is of little value. Errors may be due to emotions engendered in the patient by the test leading to an outflow of adrenaline, to over-ventilation of the lungs, or to faults in the recording apparatus.

Among other causes of an increased basal metabolic rate the leukæmias are the most important. Others are heart failure with dyspnœa, polycythemia, the active stage of acromegaly, the fevers, and pregnancy.

From the above it can be seen that a patient with a B.M.R. of 110 or 115 may well have toxicity since her estimation during health, had it been done, might well have been 85 or 90. From 90–115 is a rise equal to that of from 100–128. It is thus obvious that the B.M.R. estimation is of greater value in determining lack of toxicity, since repeated low estimations are not likely to occur in the presence of toxicity. The highest values in hyperthyroidism are rarely above 160.

The advantages of medical treatment, using such a drug as thiouracil, are that, save for severely ill patients, treatment can be ambulatory and operation avoided. The disadvantages are that it is a long treatment, that it is less certain in its effects than operation, that there are some side effects which may entail cessation, that an initial good result is frequently followed by relapse, and that during the long course of treatment required the patient often develops persistent functional features. The advantages of surgery are that there is greater assurance of relief of symptoms or "cure" and that a much shorter period of treatment is required; the drawbacks lie in an operative mortality rate and in a morbidity from certain sequelæ. The operative mortality rate, in experienced hands, has now been reduced to less than one half per cent. There is also, however, a small mortality rate under medical treatment due chiefly to thyroid crises and to heart failure from thyrocardiac disease. Choice between medical and surgical treatment will largely depend upon the attitude of the patient to an operation, on the preference of the medical adviser, and on the availability of a surgeon skilled in the specialty. Treatment with *radioactive iodine* is still on trial. While it may produce the desired effects with the least of trouble to the patient there are difficulties of dosage, and the long-term effects of such irradiation have yet to be observed in regard to genetic changes in the gonads and possible, but unlikely, local carcinogenesis. It is undoubtedly, given under highly skilled control, the best line of treatment in recurrent toxic goitre where there has been a previous operation. It is not likely to give a favourable result in nodular goitres which have become toxic.

Specific Medical Treatment. Treatment with thiouracil is usually carried out on a patient attending to her duties, household or otherwise, in more or less degree. In severe instances, however, the patient requires rest in bed and other attentions in the initial treatment. Coincident with the thiouracil administration a sedative such as phenobarbitone is given. Thiouracil itself has been displaced by methyl thiouracil and propyl thiouracil; both these are less toxic than thiouracil itself and the propyl compound a little less toxic than the methyl A dosage of 0·2 gm. thrice daily is given until the symptoms have been brought well under control, as shown chiefly by fall in pulse rate, recovery from weight loss, and general well-being. In favourable cases this is achieved in one to three months. Thereafter the dosage is reduced for a period of a few months in which the patient takes 0·2 gm only twice a day or 0·1 gm. three times. It is essential that the reduced amount should keep the patient at her improved level; any deterioration requires a stepping up to the original level. Finally a maintenance dose of 0·05 gm. is taken for some six months before entire discontinuance of the drug.

Complications of Thiouracil Treatment A host of minor, and one major, toxic reactions have followed on the administration of thiouracil and its compounds. Their incidence, when thiouracil itself is employed, has been given as about 15 per cent, but has been considerably reduced, to about 5 per cent, with methyl and propyl thiouracil. The serious reaction is agranulocytosis, which may occur even with propyl thiouracil, and with thiouracil itself had an incidence of about 2·5 per cent. About one in four of these patients died. Agranulocytosis may appear with startling rapidity and its prevention by leucocyte counts at weekly or longer intervals is therefore unsatisfactory. A more practical safeguard is to advise any patient taking the drug to discontinue it at once if any infection, particularly of the throat, occurs. Such infection follows upon the diminished resistance from the absence of white cells. Discontinuance of the drug and the immediate

harm will come, and benefit may accrue, from its removal in doubtful instances of toxicity, results do not bear this out; the functional patient usually has her symptoms made worse by operation.

The presence or absence of a palpable goitre can of course help a great deal in diagnosis. Some of the difficulties in its determination have been dealt with (p. 289), and it is there pointed out that a gland many times its normal bulk can be barely palpable. Can toxicity emanate from a gland which appears normal in size? It is difficult to say with assurance that a gland is normal in size for any one individual; a little increase in all its measurements may make it double its normal bulk. There is, as with other things, a variability in the size of the normal gland. When operation is performed for undoubted toxic goitre where the gland is apparently of normal size on clinical examination, the gland is usually found to be enlarged and, even if only slightly so, its state of vascularity leaves no doubt as to its enhanced activity.

Where the patient presents with a goitre and auricular fibrillation, it is not to be assumed that the latter is resultant upon the former. Other signs of toxicity should be sought for.

Where there still remains doubt as to the presence of toxicity a therapeutic test may be employed. While either iodine or thiouracil might be given to an ambulant patient for the purpose, the test is better made with the patient at rest in hospital. Iodine is chosen since it produces its effects more quickly but it is withheld for a week or longer, in order that a proper baseline for the study of its effects can be obtained after a period of rest and sedation. A significant gradual drop in the pulse rate, with a general amelioration of other symptoms, after a week or two of iodine therapy, will justify the diagnosis of toxicity.

Treatment of Toxic Goitre

Since the cause of toxic goitre is unknown, no treatment can be directed with confidence at the source of the disease. Ideas that the thymus initiated the thyroid changes, and that the thyroid gland was controlled by the sympathetic system, led to the performance of the operations of thymectomy and cervical sympathectomy. Both these procedures have long been discarded as unsuccessful.

As noted earlier, a proportion of patients will recover spontaneously, commonly only for a time. Such spontaneous subsidence of symptoms, a process which takes usually a year or two or more, is regarded as due to the passing off of the external stimulus to the gland.

The Alternative Forms of Treatment. The forms of treatment available are medical, surgical, and by irradiation. All are directed at the gland itself with a view to reducing its functional capability. The rationale of medical treatment is, by the use of certain drugs such as thiouracil and its compounds to prevent the synthesis of excess hormone over a period of time sufficient for the stimulus to over-production to pass off. Surgical treatment, the removal of most of the gland, leads to a similar lowering of the amount of secreted hormone to a level which corresponds to normal. Should the stimulus eventually pass off, the small remnants of gland left at operation hypertrophy, if need be, to meet the demands of the body. Treatment by irradiation, now carried out by the introduction of radioactive iodine into the body, to be taken up by the thyroid, aims at lowering, or even destroying, the function of the secreting cells. Irradiation by deep X-ray therapy is rarely successful and has now largely been abandoned.

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administration of penicillin are the sheet anchors of treatment. The injection of substances to stimulate white cell production, such as pentose nucleotide, are of doubtful value. Transfusion of packed white cells is said to be more useful.

Short of agranulocytosis a neutropenia may be observed where serial blood counts are done.

The less serious reactions are fever, skin irritations and eruptions, arthralgia, pharyngitis, conjunctivitis and enlarged salivary glands.

A non-toxic complication is gross enlargement of the gland, enough to cause pressure features. *This change is most common when large doses of thiouracil have been used.* It is obvious that, because of this risk of enlargement, thiouracil is contraindicated in retrosternal goitre.

Generally speaking, the occurrence of reactions, excepting mild ones, is an indication for cessation of treatment.

Several reports have appeared of infants being born with hyperplastic thyroid glands following prolonged taking of thiouracil by the pregnant mother. In one instance the degree of hypothyroidism was alleged to have led to defective mental and physical development. *The drug is therefore probably best given for a short period during* pregnancy. There is, however, no contraindication to subtotal thyroidectomy in pregnancy; there seems no tendency to abortion following the operation.

RESULTS OF THIOURACIL THERAPY. The presented statistical results of thiouracil therapy differ considerably, the sustained remission rate claimed by different authors varying between 25 and 80 per cent. The wide discrepancy between these figures can be somewhat explained by the early enthusiasm for a new non-operative procedure, by the zeal with which the treatment is conducted, and by the differing opinions among physicians as to the criteria of toxicity. It is now, however, generally agreed that it is unwise to give a third course of treatment where relapse has occurred twice. Operation or radioactive iodine therapy is then indicated. It is also generally held that the most successful results are obtained in patients with early diffuse toxic goitre.

OTHER SPECIFIC MEDICAL TREATMENT. It is claimed that another antithyroid drug "neo-mercazole" acts more rapidly, produces fewer complications and is generally more effective than thiouracil and its compounds. Sufficient time has not yet elapsed for the accurate assessment of its value.

Treatment with Radioactive Iodine. The indications and contraindications have already been given (p. 293). The degree of hyperactivity of the gland is assessed after giving a tracer dose of radioactive iodine by excretion tests and the use of the Geiger counter on the neck A suitable dose of I^{131} is then given orally. Improvement usually manifests itself in 6 weeks' to 3 months' time, thereafter further doses may be given as found necessary.

Surgical Treatment. Surgical treatment, subtotal thyroidectomy, entails removal of the main mass of the gland, only a small fraction being retained. Usually achieved by one operation, the procedure occasionally requires to be done in stages.

It must not be forgotten that at one time the operation was attended by such a high mortality rate that physicians were naturally averse to its performance. Successive improvements in pre- and post-operative treatment and in the technique of operation gradually revolutionized the outlook. Appreciation of these, and vigilance in their observance, are essential if a low mortality rate is to be achieved.

Briefly, the transformation has followed on the knowledge that the administration of iodine can cause considerable diminution of toxicity, on the discovery and use of anæsthetics of minimal toxicity, on the observance of hæmostasis and gentleness at operation, on the importance of pre-operative rest and sedation and on the correction of remediable deficiencies and breakdowns as far as possible before operation. Finally, the anti-thyroid drugs, thiouracil and its compounds have proved valuable in the preparation of the more seriously ill patients.

PRE-OPERATIVE TREATMENT. While other items are necessary and important in both and will be discussed, pre-operative treatments can be divided into two broad types, preparation by iodine administration and preparation by thiouracil and iodine. Briefly, in the first the patient is admitted to hospital, made to rest, given sedatives and iodine, and is operated upon when her general condition has improved, her restlessness and apprehension have been diminished, and her pulse rate has been significantly lowered. In the second the patient is given thiouracil while an outpatient, on the lines described above under medical treatment, together with mild sedative drugs, until the disease is controlled. Thereupon a time is fixed for operation and for a fortnight preceding this she is given iodine instead of thiouracil in order to diminish the great vascularity of the gland aroused by that drug. The patient is admitted to hospital a few days before the end of the fortnight's period of iodine.

The simpler "iodine preparation" is of course the older method. Its advantage is that patients can usually be made fit and safe for operation in 7–21 days; its drawback is that there is a small proportion of patients who are iodine-resistant and do not achieve the necessary improvement. The advantages of "thiouracil preparation" are that the patient is usually detoxicated to a rather greater degree than with iodine, and that there are fewer patients resistant to its beneficial effects, although, as has been said above, these beneficial effects are not necessarily lasting. It is therefore the safer form of preparation where the surgeon is not highly experienced. Its main drawbacks are that the preparation may require a matter of months, during which time the patient requires to be under out-patient supervision, that the gland does not always react to the terminal iodine in respect of vascularity and may present difficulties of technique at operation, and that the gland sometimes swells greatly to produce pressure features. There is an additional important risk that, given to patients with unrecognized "malignant" exophthalmos (p. 306) with mild toxicity, thiouracil may make the proptosis of the eyes markedly and seriously greater in degree.

The choice between the two lines of pre-operative treatment is an individual one for the surgeon. Some favour thiouracil treatment as routine; others, especially those with long experience before the discovery of thiouracil, rely on iodine alone for almost all patients, reserving thiouracil for the most severe types or for those apparently resistant to iodine. In the latter event thiouracil is unlikely to have any effect for a much longer period after iodine has been taken than would otherwise be the case. It is then better to give no drug at all for a period of a month or more between iodine and thiouracil therapy.

The routine of the average patient who is admitted to hospital with a view to operation after iodine preparation is as follows. The patient is made to rest in bed but is allowed up to the bathroom. A fruit drink, to which a spoonful of glucose is added to every tumblerful, is made freely available by the bedside; the patient is encouraged to

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drink at least a pint of this daily. Such an amount of fluid should not, of course, be offered to anyone with heart failure. Sedative drugs, such as phenobarbitone gr. ½–1 b.d. are given. The best sedative, however, is probably not a drug but the reassurance obtained by the patient from the presence of several patients in the same ward recovering from the operation and the disease. This, and their realization that the nursing staff is experienced and sympathetic, make a wealth of difference to the apprehensive patient. Such an affirmation is not to be taken as advocating a special ward or clinic for these patients; on the contrary they are, from all points of view, better regarded as ordinary surgical patients and mixed with them. Segregation, either in special wards, rooms, or even by screens, only heightens their apprehension.

The patient's weight is noted on admission and a general examination carried out together with any special investigations relevant to the particular patient. Ordinarily in the first few days the pulse rate falls and steadies to a level which can be regarded as a true base line from which to measure the effects of the iodine. A preparation of iodine is then given until the pulse has fallen significantly and the patient's nervousness and restlessness have abated. The iodine may be given in several forms. The most popular is the preparation known as Lugol's iodine, a 5 per cent solution of iodine in 10 per cent potassium iodide. Although divided and increasing daily doses of this have been advocated, there is no advantage in such methods over a single daily dose of 10 minims, taken in milk. Lugol's iodine is somewhat unpleasant of taste and is no more efficacious than the more palatable sodium iodide, given as 5 grains thrice daily in solution or in tablet form. Depending upon the severity of the condition the iodine will require to be given for some 7 or 21 days. The optimum time for operation, considering the pulse alone (which is insufficient) will be when the rate is steady in the seventies or eighties. But, if in a patient who is cool and calm and not emaciated the pulse will not fall to these levels, operation should be proceeded with even if the pulse be in the nineties. The safeguard is to restrict the scope of the operation to subtotal removal of one lobe if the pulse rate keeps rising while that lobe is being mobilized and resected, or if the rate should get to a high level such as 130 or more and remain there. The other lobe is resected a week or two later.

In a few patients there is a "resistance" to iodine. The general beneficial results do not follow its administration. But in some the "resistance" seems limited to the pulse rate only, all other features being markedly bettered. In these, with the safeguard that the two-stage operation may have to be adopted as described above, too much regard should not be paid to the pulse rate.

Such routines are suitable and sufficient for 95 per cent of patients. The other 5 per cent may require special attention. Where there is extreme restlessness, stronger sedatives, even hyoscine, will be required. Great emaciation will entail a longer preparation and careful dieting Patients with auricular fibrillation should be given digitalis along with iodine: the digitalis should be withdrawn three days after operation unless there is indication for its continuation. Quinidine should not be given pre-operatively.

Principles of Operative Treatment

Anæsthesia. The use of anæsthetic agents of such toxicity as ether and chloroform is strongly contraindicated in operations for toxic goitre. The patient should have her nervousness allayed, arrive in the theatre asleep, and be kept in a light plane of anæsthesia throughout. Pre-medication with omnopon-scopolamine, the administration of a

basal hypnotic such as bromethol ("avertin"), followed by nitrous oxide and oxygen during the actual operation, achieve this end. Where such routine proves insufficient it may be supplemented by fractional doses of pentothal injected into a vein in the foot. Cyanosis must be prevented throughout. Most surgeons and anæsthetists favour intubation of the larynx and trachea with a single large tube; a few object. The various manipulations of the operator in dislocating the lobes lead to the trachea being compressed or kinked. This in turn leads to an anoxæmia which must be avoided; the presence of an indwelling airway helps in this.

Opponents of intubation say that it requires a deeper degree of anæsthesia to introduce the tube than is desirable; this is denied by those expert in its use. Again, these opponents emphasize trauma to the larynx and trachea by the indwelling tube; this objection is more theoretical than real if the tube is well lubricated and passed with skill.

Whether a tube be employed or not, a clear airway must be maintained although in patients with large glands there may have to be some temporary partial obstruction in mobilizing a lobe. It is essential that any dyspnœa caused by this should be of short duration at any one time.

General anæsthesia may be supplemented by local infiltration of the neck with some 50–100 ml. of 0·25 per cent solution of procaine, to which adrenalin 1 in 1000, is added, 11 minims to the 100 ml., producing a concentration of 1 in 150,000 adrenaline. This is best introduced firstly subcutaneously throughout the line of the incision and then under the platysma over the area in which the flaps will be reflected. The introduction of the fluid also facilitates dissection of the flaps, and the adrenalin, in addition to enhancing the local effect of the procaine, produces a constriction of the small vessels and a bloodless field. Many surgeons, including the author, infiltrate the neck thus, but, instead of using the adrenalin solution described, employ normal saline instead of procaine solution and add adrenaline in the same concentration. With skilled anæsthetists the use of procaine is unnecessary. But hæmostasis by the widely infiltrated adrenalin is greatly helpful.

While the operation may be done entirely under local anæsthesia, such a procedure imposes a strain on the toxic thyroid patient that is nowadays unjustifiable. Additionally it entails a more prolonged operation and a strain on the surgeon himself. Where it has been practised, there is a choice between cervical plexus block and infiltration methods.

The Operation. Details of operative procedure should be studied elsewhere. It is proposed here only to emphasize certain principles and discuss problems that may arise.

Whatever may be the real cause of a post-operative thyroid crisis, it is best from the practical point of view at operation to regard it as due to shock in a patient highly sensitive to shock Loss of blood and rough handling must therefore be studiously avoided. During the initial dissection it should be assumed that some blood loss will be incurred during actual section of the gland and every effort made to limit the loss to what happens then. If the neck be infiltrated with the saline-adrenalin solution mentioned above, and if the anterior jugular veins are carefully avoided, or ligated previously if the muscles are divided, then the gland should be reached with but a trifle of blood loss.

The line of the incision should be carefully marked out with Bonney's Blue before infiltration; otherwise the natural creases may be difficult to define.

Whether the infrahyoid muscles should be divided transversely or not will chiefly depend upon the size of the lobes and upon the skill, experience and personal predilections

of the operator. The step should not be omitted if it is going to make for significantly more gentle manipulations. With increasing experience the surgeon will tend to divide the muscles less frequently and will realize that it is the consistency of the gland much more than its bulk that demands the step. Quite large glands, if they are soft and pliable, can be rolled out easily from their recesses without transverse division of muscles; smaller, hard, unyielding glands may entail the division.

As to how much gland should be removed and how many main vessels tied, the problem is how to be sufficiently radical and yet not produce a permanent hypothyroid state. The difficulty is to a large extent solved by the ability of the gland to regenerate up to a point. The aim should be to produce in the patient a temporary mild subthyroid condition which, with hypertrophy of the remaining tissue, will in a few months pass to the euthyroid state.

The problem is somewhat different in diffuse and nodular goitres. To deal with the diffuse goitre first, it is meaningless to say, as is often said, that seven-eighths or nine-tenths of the gland should be removed. What should be done is to leave a portion of each lobe on either side of the trachea roughly the size of a normal thyroid, perhaps less, perhaps more. The degree of difference should be assessed in relation to the age of the patient, the degree of toxicity, and as to whether the inferior arteries are or are not ligated in continuity. As to deprivation of blood supply, it is essential in diffuse goitre, in order to mobilize each lobe thoroughly, to divide the superior vessels. Because additional unnamed smaller vessels reach the toxic gland from other sources, the inferior arteries may also be occluded without depriving the gland of all blood supply. (The inferior artery is not usually dealt with by division and ligature of both ends. It is most commonly ligated in continuity, this being as effective and a safer method in dealing with a vessel which is deeply placed and intimately related to important structures.) Some surgeons ligate the inferior artery in all instances. The writer does not practise this in elderly people, unless they are severely toxic, since there is a greater risk of subsequent myxœdema than in the young. In a young patient with much toxicity the gland should be radically resected, little more than half a cubic inch of each lobe being left, and all main vessels should be occluded.

Little in the way of guidance can be given as to the extent of resection of toxic nodular goitre since it will appear at operation in some that the toxicity must be originating in the tissue around and between the nodules, so degenerate are the latter, while in others the nodules would seem to be composed of functional tissue. But the external appearances are often not helpful. Subtotal lobectomy should include all obvious nodules but occasionally the operator in dealing with lobes riddled with nodules may find himself leaving tissue which is doubtful as to function and which may contain the seedlings of further nodules. A preliminary survey of both lobes before resection is necessary. It may be found advisable, since nodules have a tendency to develop more in the lower parts of the lobes, not to divide the superior vessels on one or other side or to dislocate and remove the superior poles. Instead, the portions below these are entirely, or almost entirely, removed, and the inferior arteries tied, thus leaving only a little normal, possibly overactive tissue, at the superior pole. Where the nodular process affects one lobe only, the other lobe may be found normal or enlarged diffusely; in the latter instance it must be subtotally resected. Clearance of any obstructive features is of course as necessary in toxic as in non-toxic patients.

In all operations the thought of possible injury to the recurrent laryngeal nerves should be borne in mind. Most surgeons do not seek particularly to expose the nerve but aim at avoiding its injury by leaving untouched that part of the lobe, near the entrance of the branches of the inferior artery, which is in the most intimate relationship, by gentle handling of the lobe, and by tying the inferior artery well out from the gland. Lahey (Lahey, F. H. (1944) *Surg. Gynec. Obstet.* 78, 239) advocates a technique in which the nerves are deliberately exposed, and the parathyroids identified, before resection. This entails wide transverse division of the infrahyoid muscles. The criticisms of this method are that the nerve is as likely to be injured in the search for it as when it is left unidentified and that the additional time spent in seeking the parathyroids is unjustified since post-operative tetany is such a rare event.

Post-operative Treatment. The immediate post-operative essentials are that the patient shall be kept quiet and cool and have her fluid loss made good. For the last, unless loss of blood at operation is severe enough to necessitate transfusion, two rectal infusions, at an interval of 8 hours, of a pint of saline with 5 per cent glucose, will be adequate. Sedation will necessitate the giving of morphia in the first 48 hours, after which milder remedies will suffice. Further iodine medication should not be given. Excessive warmth will help any tendency towards a crisis; few bedclothes should be allowed; fans and even cold sponging may be needed in hot weather. The patient should be propped up in bed as soon as anæsthesia has been completely recovered from. Some degree of tracheitis and dysphagia almost always follow thyroidectomy. A warm mixture of sodium bicarbonate and a linctus are useful in this connection. The treatment of a post-operative crisis is discussed on p. 301. A moderate rise of temperature and a rather higher rise in pulse rate is to be expected, reaching their maximum usually on the second day and then declining. It is rare to have any anxiety for the patient after the lapse of 48 hours. The skin stitches and drains should be removed at the end of 48 hours.

Auricular fibrillation, where such was not present pre-operatively, need rarely cause alarm where other features suggesting a crisis are absent. Where the rate becomes very high, and this will be more common in previously established fibrillation, digitalis should be given or maintained for a few days. When fibrillation does not cease by the tenth day, quinidine sulphate, 5 grains thrice daily, should be given for 10 days. Should it not then have ceased it is unlikely that further quinidine will be of benefit, although the fibrillation may later be found to have stopped.

If the full value of the operation is to be obtained a good convalescence is essential. The nervous strain of the illness and of the operation entail that the patient should have a fair period of rest before resuming household or other duties. If such is not given the patient may have an early partial relapse or, more commonly, develop troublesome functional features. As far as is possible, a married working-class woman patient should remain in hospital for a fortnight after operation and then have at least 3 weeks in a convalescent home. Thereafter for some 3 months she should endeavour to have an afternoon rest period.

The Complications and Sequelæ of Operative Treatment

Hæmorrhage. Hæmorrhage following operation is of importance on two counts. Firstly, if of significant amount, it will itself induce the shock which it is imperative to avoid if crises are to be prevented; if the bleeding does not abate in a reasonable time,

then the further anæsthesia and exploration required to arrest it will induce an even greater degree of shock. Secondly, if the blood does not escape in sufficient amount through drainage tubes the neck becomes swollen with a mass of blood and asphyxia ensues. It is difficult to believe that the pressure of the retained blood could compress the trachea and the asphyxia is more likely due to œdema of the cords and larynx. A suggestion is that the pressure of the extravasation causes collapse of the internal jugular veins, which in turn produces a local rise in the venous pressure where a considerable shutting off of the venous return from the larynx has already occurred from the operation, and œdema ensues. Since the asphyxia may come about suddenly, it should be a routine that sterilized scissors and dissecting forceps are available at the bedside; rapid release of pressure can then be obtained by opening up the wound.

Careful hæmostasis during operation should make this complication a rare one. While the trend is towards less frequent drainage in thyroidectomy, the operator will do well to elaborate such technique in non-toxic patients before embarking on its performance in the more vascular toxic ones. Where drains are inserted, care should be taken that close stitching does not frustrate their purpose.

The more serious hæmorrhages originate by slipping of a ligature from the divided superior artery, from a like occurrence on branches of the inferior artery in the gland substance if the main trunk has not been ligated, less commonly from small arteries in the infrahyoid muscles and in the edges of the skin wound. It is unnecessary to re-open the wound in the lesser degrees of the complication even if the neck be somewhat bulged; in these the blood either becomes absorbed or, later on, the scar gradually thins at one point and separates to allow the serum to exude. In neither instance is the ultimate appearance of the scar affected. Probing to release such collections should be avoided as likely to introduce infection. The wound will require exploration where blood loss has been more severe; blood transfusion may then be necessary.

THE THYROID CRISIS OR STORM. This most dreaded of all complications may be entered upon immediately after operation, the patient never really regaining consciousness, or it may supervene at any time in the next few days. Its appearance, however, after the lapse of 48 hours from operation is rare and is then possibly associated with severe infection in the field of operation. Typically in a crisis the patient becomes restless, delirious and cyanotic; the pulse mounts until it becomes all but uncountable; the respirations likewise increase in rate to 50–60 per minute, the skin is burning hot and dry, and the temperature rises to 105–106°F or more; unconsciousness ensues and the patient usually dies in a few hours. Restlessness and delirium may, however, be absent. While this is the commoner form of crisis, there is another type to which the qualifying term "apathetic" has been suitably attached. In this the patient pre-operatively is quite disinterested in her illness in which characteristically there is much wasting and profound muscular weakness After operation there is the same mental apathy and prostration and the patient dies quietly on the first or second day with but relatively moderate rises in temperature and pulse although the respiration rate rises much more significantly.

It may be interpolated here that such crises are not necessarily associated with operation. They may occur spontaneously, or be excited by infections and fevers, especially in those who have emaciation, uncompensated heart disease and psychoses as the result of chronic severe thyrotoxicosis. Crises have also followed upon surgical interference not

directed towards the goitre, such as the removal of teeth or tonsils, in patients in whom toxicity is marked.

Some patients exhibit severe post-operative reactions as to pulse rate and temperature without the other features of a crisis and there is argument as to whether these are lesser degrees of the state or have no relation. There is commonly a moderate rise of temperature and pulse after every goitre operation. Occasionally the pulse may, during the first 48 hours, rise quickly to 140 or more, or auricular fibrillation may occur; yet there is no cyanosis or disorientation, the temperature rises only to 101–102°F, the respiration rate but slightly, and restlessness is easily controlled. The treatment of such patients causes little difficulty to an experienced nursing staff.

Where, however, a real crisis develops, the problem is an entirely different one, so difficult and apparently as yet insoluble that every emphasis must be laid upon its prevention by pre-operative measures. Active treatment is devoted towards controlling the anoxæmia by the giving of oxygen, to reduction of the hyperpyrexia by cold sponging, and to sedation where restlessness is a feature. In accordance with various theories many additional treatments have been advocated such as intravenous saline and glucose, with perhaps the addition of Lugol's iodine, thyroid extract, anti-thyroid drugs, and adreno-cortical hormone. So rare is recovery that it is impossible to assess the respective merits of these various measures.

Other Causes of Post-operative Death. Deaths other than from hæmorrhage and thyrotoxis crisis may occur and, though now uncommon, are chiefly associated with longstanding thyrotoxic heart disease where pre-operative measures have not been altogether successful. Sudden arrest of the heart or fatal pulmonary embolus may then happen; more commonly death would seem to occur slowly and quietly from bronchopneumonia several days after operation. It is possible that this last type is a subacute form of "apathetic" crisis.

Tetany. Considering the radical nature of the operations performed and the fact that most surgeons, including many of the most experienced in this work, do not go out of their way to identify and preserve the parathyroid glandules, tetany is a rare complication. One or two of the glandules may quite often be found in the removed specimen. It would seem that tetany will not occur if even only one glandule is retained.

Four glandules are commonly and "normally" sited in relation to the posterior aspects of the superior poles and to the posterior aspects of the lobes lower down near the entrances of the inferior arteries or behind the inferior poles. But anomalies in their positions are common (see p. 249) and it would seem as if the superior glandules, the ones most likely to be removed at operation, are usually cleared off the surface of the thyroid in any dissection which keeps close to the capsule in freeing the superior pole. It is of note that one of the objections, particularly on the part of continental surgeons, to ligature of the inferior thyroid arteries, is that the blood supply to the parathyroid glandules will be affected. Experience does not confirm this fear; the collateral circulation is adequate. None the less, the surgeon who does not try to identify the parathyroids, an ability to do which necessitates some practice, must be careful and gentle in extricating the thyroid from surrounding tissues. Some glandules, however, are so neatly embedded in crevices on the thyroid surface as to escape detection and their removal cannot be avoided.

Identification of the glandules, therefore, is desirable although undue time should not be expended in the process. They vary in size and shape, chiefly according to the pressure

exerted on them by the enlarged thyroid. Typically they are rather ovoid, their long axis measuring rather more than half a centimetre, their shorter half of this again, but by compression they may be quite flattened. In colour they vary from yellow to a light café-au-lait. They have chiefly to be distinguished from small lobules of fat. A characteristic of a parathyroid glandule is that slight trauma will produce a subcapsular hæmorrhage.

Post-operative tetany is either mild or severe. In the former, which does not appear until the second or third day or later, it passes off spontaneously, and its features can till then be ameliorated by the drinking of a glass of milk or two daily. It is probably due to the removal of some of the parathyroids with either transient damage, such as by hæmorrhage or œdema, to the remaining ones. The symptoms of the mild form do not usually go beyond the "pins and needles" feelings although positive Chvostek and Trousseau's signs may be elicited.

The severe form, fortunately rare, is a permanent condition where all the glandules have either been removed or irremediably damaged. The symptoms may appear on the day following operation or be delayed for a few weeks. Numbness and tingling in the extremities is followed by carpopedal spasms and cramps. The patient is markedly apprehensive. Laryngeal spasms with respiratory obstruction may follow; death has occurred from such but is unlikely. The initial treatment in the severe form is the intramuscular administration of parathormone and the intravenous administration of calcium salts. An indifference to parathormone develops and its permanent administration is therefore not of value. Long-term treatment is directed toward maintaining the serum calcium at a normal level by the giving of large doses of calcium lactate by mouth and reducing the phosphorus intake. The omission of milk, which is rich in calcium but also in phosphorus, from the diet, or reduction in its intake, is helpful.

Recurrent Laryngeal Nerve Palsies. The intimate association of the inferior or recurrent laryngeal nerve to the thyroid (p. 249) renders it particularly liable to injury during operation, by stretching, bruising, division, or inclusion in a ligature. There are two schools of thought as to the attitude to be taken toward the nerve during operation. One holds that the nerve should always be clearly exposed before the lobe is resected; the other holds that such dissection is as likely to cause as to avoid trauma to the nerve and that with careful and gentle dissection in the right plane, bearing in mind the probable situation of the nerve, injuries are as infrequent. Deliberate exposure of the nerves certainly adds to the time of operation. Traction injuries are probably commonest and result from rough mobilization of a lobe.

The recurrent nerve supplies all the laryngeal muscles except the cricothyroid, which is the tensor of the vocal cords and is supplied by the superior laryngeal nerve, and part of the arytenoideus muscle. Its complete division results in the corresponding cord becoming immobile and assuming the cadaveric position, halfway between adduction and abduction. Where both nerves are completely divided and thus both cords are incapable of either adduction or abduction then phonation is lost except that the false cords may gradually develop to afford some assistance. Much more commonly, fortunately, only one nerve is damaged and the opposite cord compensates by over-adducting to meet the immobile one Phonation is thus possible and gradually becomes stronger, although, because of the difference of tension in the two cords there is a rather hoarse voice.

In incomplete lesions it is the abductor element that is paralysed and then usually temporarily. The cord lies fixed in the midline, and naturally, if the occurrence be bilateral, there is great dyspnœa and stridor, with commonly the necessity for tracheotomy. Where only one side is affected there is little discomfort beyond some dyspnœa on exertion. Where after several weeks of tracheotomy there is no return of function in the bilateral cases, operations to prevent the necessity for permanent tracheotomy are called for. These aim at producing an interval between the cords. King's operation consists in shifting the position of the arytenoid laterally. Kelly resects the arytenoid through a window in the thyroid cartilage and stitches the cord to the fascia outside the window.

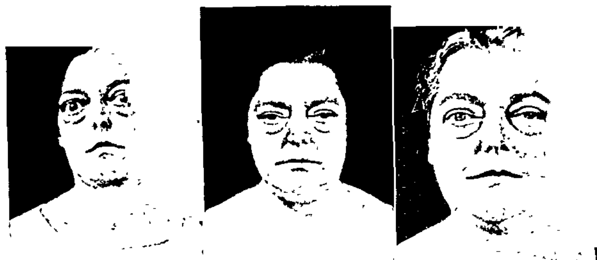


FIG. 149. Diffuse toxic goitre with auricular fibrillation treated by subtotal thyroidectomy The middle photograph shows the same patient 10 years later, the war intervening, with well marked myxœdema The photograph on the left is the same patient after treatment with thyroid extract.

It should not be forgotten that post-operative aphonia is frequently a functional condition. In this the voice is often reduced to a whisper but little or no dyspnœa is present. Such a condition may present immediately after operation or appear suddenly during the next few days. Laryngoscopy shows that in adduction of the cords there is slight impairment of their complete approximation. There is usually spontaneous improvement although in some patients re-education by exercises may be necessary.

POST-OPERATIVE MYXŒDEMA. The transient sub-thyroid state aimed at by resections for toxic goitre (p. 298) is chiefly characterized by a mild increase in weight beyond that which the patient is accustomed to or to care for; there is generally a spontaneous corrective adjustment after a period of some six months or a year.

Myxœdema proper, when it follows upon operation, is most likely to occur where too radical an operation has been performed on a gland mistakenly thought to be producing toxicity. But even then it is uncommon. Where it has occurred in undoubted thyrotoxicosis it has sometimes been attributed to the prolonged administration of iodine after operation, this tending to prevent compensatory hyperplasia, but even without this it does occur occasionally, particularly in older subjects and in them the writer tends to avoid ligation of all main arteries. It would seem (see Fig. 149) that where there has obviously been severe toxicity at one time, the disease may pass to a smouldering stage,

and over-radical procedures may then result in myxœdema. Auricular fibrillation is more associated with toxic nodular than with toxic diffuse goitre and it may well be that in the remnants there is more in the way of degenerative nodules than of functioning thyroid tissue.

It is alleged that myxœdema is more common where thiouracil has been given as pre-operative medication; since thiouracil by itself does sometimes produce myxœdema it is possible that this is so. Considering the variable effects of thiouracil it is certainly more difficult to estimate how much gland to leave behind than when iodine is used.



FIG. 150 Diffuse toxic goitre in male age 27 showing complete recovery from lid retraction in 15 months after subtotal thyroidectomy.

While myxœdema is a complication that can be amended by the use of thyroid extract, and while the surgeon may have a fairly wide margin within which to work, it is a state to be avoided by the exercise of judgement in the individual patient. The daily ingestion of the extract can be a nuisance and an expense and, additionally, the actual optimum amount for a particular patient may require quite a little adjustment. Finally, in this connection, there seems no evidence that apparently more refined products have any greater efficiency than the dry thyroid extract of the pharmacopœia.

RESULTS OF OPERATION: RESIDUAL AND RECURRENT THYROTOXICOSIS. Ten days from the date of operation it should usually be possible to tell where iodine only and not thiouracil, has been used in the preparation, how much benefit is likely to accrue from the operation. The pulse should then usually be normal in rate while the patient is at rest, there should be an absence of irritability and fidgetiness, the patient should express a feeling of coolness or even of cold. Where there has been auricular fibrillation the pulse in some 50–60 per cent of cases will have become normal in rhythm; in others it will fibrillate more slowly. Provided the convalescence be conducted properly, in three months, time there should have occurred a significant gain in weight; the pulse rate, while still a little volatile, should be normal at rest, the hands should be cool and dry; tremor should be absent or minimal; lid retraction should have gone or be considerably

improved, but true exophthalmos will take much longer to lessen or disappear, frequently never entirely clearing up; the patient should express a definite betterment in all ways.

Where such benefits have not been obtained then the operation has not been sufficiently radical and there is *residual* thyrotoxicosis. This sequel is most likely in young patients operated upon by as yet inexperienced surgeons who are naturally over-cautious. Thiouracil will usually cause suppression of this residual toxicity in a permanent fashion.

In *recurrent* thyrotoxicosis, on the other hand, there is a reappearance of symptoms after their complete remission following operation for a period of usually many years. Presumably the original stimulus from the hypothalamus abated and then reactivated, to find sufficient gland tissue for hypertrophy to produce toxicity. The onset of recurrent toxicity seems often to be associated with a further emotional stress. Its incidence is about 5 per cent. Secondary operations on the gland should if possible be avoided since the technical difficulties may be great on account of scar tissue. Thiouracil should first be tried; it is often successful. If radioactive iodine is available and can be expertly given, it will serve even better. Where further operation is to be performed, access to the enlarged lobes, after reflecting the flaps, is best obtained by incisions along the anterior border of each sternomastoid muscle. Before any such secondary operation is embarked upon, the state of the vocal cords must be determined, since unnoticed damage may have occurred to these at the primary operation. Knowledge of such damage will warn the operator as to exercising great care on any particular side; it may even cause him to abandon his project.

"MALIGNANT EXOPHTHALMOS"

(Thyrotropic Exophthalmos: Exophthalmic Ophthalmoplegia: Hyper-ophthalmopathic Graves' Disease)

This is a morbid condition of unknown cause characterized by severe protrusion of the eyes, often associated with ocular muscle weaknesses or even paralyses, and associated or unassociated with thyrotoxicity. The protrusion may reach a degree in which the eyelids fail on attempted closure to cover a large portion of the protruded globe; consequent corneal ulceration may be so severe as to lead to loss of the eye. The degree of protrusion bears no relationship to the amount of thyrotoxicity, if the latter be present. Of the several synonymous names given to the condition possibly the best is *malignant exophthalmos*. Although it may wrongly suggest neoplastic disease, the term is impressive of the need for recognition of a state of grave importance.

The condition is one of the enigmas of endocrinology and its consideration first leads to a discussion of the theories of the causation of exophthalmos in toxic goitre.

The term *exophthalmos* is used loosely in clinical descriptions and thus is frequently made to be inclusive of the upper *lid retraction* which is a common eye feature in toxic goitre. The two conditions are distinct. Exophthalmos is synonymous with proptosis, an abnormal forward protrusion of the eyeball; lid retraction signifies a drawing up of the upper lid. The two conditions may exist separately or together. Lid retraction is generally regarded as due to a spasm of the levator palpebræ superioris, and is most recognizable by the margin of sclera visible above the iris; in most people the margins of the lids cross the upper and lower parts of the circumference of the iris. In exophthalmos the lids naturally become more separated but this is initially roughly made more manifest

by the sclera becoming evident between the lower lid and the iris, although, as the proptosis increases, a band of sclera is visible both above and below the iris. Exophthalmos and lid retraction may both be unilateral or of unequal degree in the two eyes.

The degree of exophthalmos can be measured by the exophthalmometer, which, applied to the sides of the forehead, has a scale in millimetres by which the protrusion can be measured. The drawback of such readings is that there is a fair variation in the setting of normal eyes and they are really of more value in the determination of changes after the first reading.

The causation of true exophthalmos is still obscure. It is now largely agreed that it is the increased bulk of orbital contents which pushes the eyes forward; what remains unsolved is what produces the intraorbital change. There is a great increase in the amount of fatty tissue within the orbits, not only in the ordinary fibro-fatty tissue but within the extrinsic ocular muscles. It has been possible, during orbital decompression operations designed to relieve extreme and dangerous exophthalmos, to examine the orbital contents during life. There has then been found enormous swelling of the extrinsic eye muscles, chiefly from œdema, with a similar œdematous condition of the increased fat. Whether or not other agencies participate it would seem that the increased bulk of the muscles and fat plays the major part in pushing the globe forward.



FIG. 151. Malignant exophthalmos in male aged 54 Nine months history of diplopia especially on looking to the right. Weakness of both external recti muscles, right more marked than left. Subtotal thyroidectomy resulted in no improvement in the ocular signs, and irradiation of the pituitary gland also failed

What initiates these changes? The degree of exophthalmos in toxic goitre bears no relationship to the degree of toxicity. Indeed true exophthalmos may be absent in marked instances of diffuse toxic goitre and its presence in toxic nodular goitre is exceptional. Malignant exophthalmos may occur in the absence of toxicity although a goitre is usually palpable; generally speaking, however, there is associated some undoubted toxicity. The natural first conclusion that thyrotropic hormone produces both thyroid over-function and exophthalmos is therefore unsatisfactory. It is true that some thyroidectomized animals have become exophthalmic after the injection of thyrotropic hormone. Likewise a wide variety of animals, with or without their thyroids, it is claimed, have become exophthalmic after injection of anterior pituitary extracts. Findings in experimental animals, however, are as likely to be as inapplicable to humans in respect of exophthalmos as elsewhere. It may be that there is an as yet unidentified element in the anterior pituitary secretion which is responsible for exophthalmos.

Whether *malignant exophthalmos* is a more pronounced result of whatever causes ordinary degrees of exophthalmos or whether a different or additional force comes into play is a subject for conjecture. The practical importance of the matter is the recognition of severe exophthalmos which is associated with slight or no thyrotoxicity, for then operation or, even worse, the administration of thiouracil, may worsen the condition

profoundly. Where, in addition, as may happen, the more advanced features of *malignant exophthalmos* appear, with swelling and bulging of the lids, chemosis of the conjunctivæ, and weakness or paralysis of ocular movements, the warning is intensified. The commonest paresis or paralysis affects upward movement of the eye; loss of abduction and then of adduction are the next most frequent; downward movement is rarely affected. In the earlier doubtful stage of the condition it is well to remember that males are more liable than females.

The treatment of *malignant exophthalmos* is difficult. Fortunately in most instances the proptosis stops at a point where the risk to the eyes and to the sight is slight. In some there is a degree of recession after a time; in others, in spite of or because of treatment, it progresses until ulceration of the cornea occurs. Where the thyrotoxic features are undoubted and well marked, subtotal thyroidectomy often causes improvement of the proptosis and paresis; it is wise to give such patients a course of thyroid extract for a period after operation. Thiouracil, however, should never be given, even where much thyrotoxicity is present, either as medical treatment or as preparation for operation. Whether it is that it temporarily knocks out the thyroid more completely than does subtotal thyroidectomy or acts in some other way, it has been followed by worse results than has operation. Where the condition has become worse after operation or after thiouracil it can be argued that the condition might have progressed if neither therapy had been given. The impression is, however, that subtotal thyroidectomy, where little or no thyrotoxicity is present, causes exacerbation of the exophthalmos and that thiouracil, however much toxicity be present, is even more culpable.

Various treatments have been given on theoretical grounds. None have consistently produced an amelioration of proptosis; each has had its apparent occasional success. Thyroid extract and stilbœstrol, as antagonists of thyrotropic hormone, have been much employed. The pituitary has been irradiated in order to diminish its activity. Irradiation has also been directed at the orbital contents.

Where corneal ulceration is threatened the eyelids may require to be sutured together, a tarsorrhaphy, for a period. Where this fails and the condition is progressive, decompression of the orbit, Naffziger's operation, is indicated. In this, after making a frontal bone flap, the operator raises the frontal lobe of the brain and removes the roof and lateral wall of the orbit. The orbital fascia is then incised. Choked disc is another indication for such a decompression, the results of which are highly satisfactory. Enucleation of the eye may be necessary where infection is present or threatened and when sight is lost.

Malignant Goitre

Secondary deposits from malignant neoplasms originating elsewhere are mainly of academic interest since they are rare, usually small and apparent only at necropsy, and seldom evident before the diagnosis of the primary neoplasm has been made for other reasons.

The important form of malignant goitre is the primary malignant tumour. The subject is a complex and difficult one from all aspects, clinical, pathological and therapeutic. Thyroid neoplasms may behave in a bizarre manner quite at variance with the behaviour of malignant tumours elsewhere; they have been classified in many different ways but never satisfactorily; the histological criteria of malignancy vary with different

observers; the most efficient lines of treatment have yet to be found. Generally speaking, the outlook is grave.

Ætiology. There is much difference of opinion as to the prevalence of the disease. The points mostly in dispute are the commonness or otherwise of its supervention upon simple nodular goitre and the criteria of malignancy in histological examinations. These matters have considerable practical importance.

Numerous papers with statistical figures have been published on these points. The statistics presented by different writers are not often really comparable, show many discrepancies, and rarely take a broad enough view of the subject. Almost all the figures given relate the number of malignant goitres found to the total number of goitre operations performed, without reference to patients not operated upon, let alone reference to people with goitres who seek no advice and indeed may not know they possess a goitre. Thus they are selected series and the real incidence of malignancy is probably much less than the published figures suggest.

Different writers do not divide their material in a uniform way, but from an analysis of many papers certain figures emerge:

1. In large series of operations for all types of goitre, toxic and non-toxic, diffuse or nodular, the occurrence of carcinoma is given variously as from 1–3 per cent. The malignant state may have been diagnosed or suspected before operation or it may have been an operative or even only a histological finding.

2. If diffuse non-nodular goitres are omitted from the total operation figures, then the incidence of carcinoma in nodular goitres, toxic and non-toxic, is variously given as 4–8 per cent. The discarded smooth goitres may be regarded as almost entirely toxic diffuse goitres in which concurrent carcinoma is a great rarity.

3. If only non-toxic nodular goitres, whether the nodules be multiple or single, be considered, then the figures given for carcinoma lie between 1–17 per cent. Much variation is only to be expected in this item since the diagnosis of toxicity varies with the clinician.

4. If the figures of the non-toxic nodular goitres are further subdivided as to whether the nodules were single or multiple, then

(*a*) in multinodular goitres the incidences of malignancy given lie between 4–10 per cent;

(*b*) in goitres with a single (or apparently single) nodule, the incidences given vary from 4–24 per cent; most authors incline to the higher figure.

Such high incidences have led in recent years to strong advocacy of the removal of nodular goitres, particularly those with single nodules, as prophylaxis against the supervention of malignancy. But, whether the removal of these nodules is or is not advisable, the high incidences cannot be allowed to stand unchallenged. The arguments against them are:

1. Established carcinoma of the thyroid, undisputed from both clinical and pathological aspects, is an uncommon disease. In 1939 in England and Wales only 300 people died of carcinoma of the thyroid, i.e. approximately 1 in 1600 deaths. At the London Hospital, with its 900 beds and a radiotherapy department drawing patients from a wide region, the average number per year for 7 years, 1945–51 was 6. Yet nodular goitre is a common condition, much commoner than is appreciated clinically; small single or multiple nodules of hyperplasia in the thyroid are among the commonest casual findings in necropsies.

2. The diagnosis in the various series referred to above was based in a majority of cases upon a histological finding only, without real clinical confirmation. Here, again, is an explanation of the varying figures, for histologists differ much in their criteria of goitre malignancy. The pitfalls of histological diagnosis are elaborated later (p. 314). But, apart from this, very few of the patients regarded as having carcinoma went on to produce metastases after removal of their tumour, far too few considering the known tendencies of malignant disease.

While it is likely that the higher figures in these incidences are exaggerations, this does not rule out the contention that malignant change supervenes more frequently upon a simple goitre than upon a normal gland. Although this conclusion has been disputed it receives fairly general acceptance because of an undoubted greater incidence in endemic areas.

The disease may occur at any age but is far more common in middle life and onwards. Its incidence in children is probably very low indeed despite suggestions to the contrary, suggestions again mainly based on histological opinions. There has been only one undisputed instance of malignant thyroid tumour at the Hospital for Sick Children, Great Ormond Street, in 30 years.

The sex ratio is variously given as between 8 to 1 and 2 to 1, females to males. Most series give the ratio about 4 to 1. About half of the patients give a history of a pre-existing goitre; not all pre-existing goitres, however, are evident.

Classification. The primary malignant tumours of the thyroid may be classified thus:

1. Carcinoma
   - (*a*) Adenocarcinoma
     - Papillary
     - Follicular (or synonyms, Colloid, Vesicular Acinar, Alveolar)
     - Solid
     - Hürthle-celled
   - (*b*) Undifferentiated Carcinoma
     - Small spheroidal-celled
     - Giant-celled
     - Spindle-celled (Carcinoma sarcomatoides)
   - (*c*) Squamous-celled (epidermoid) Carcinoma
2. Sarcoma
   - (*a*) Fibrosarcoma
   - (*b*) Reticulum-celled sarcoma

Of these the carcinomata are by far the commonest and most important. The synonyms for "follicular" are used widely by different writers, who again often use them alternately in the same article in a way apt to confuse.

Many classifications have been presented and it must be admitted that the one given above, like all morphological classifications of carcinoma of other tissues, is far from satisfactory because in many of these growths two or more types of cancer tissue are mixed. Thus in a growth composed mainly of follicles there may be areas in which the cancer cells are in solid groups, and vice versa. In a follicular or solid adenocarcinoma there may be areas of spindle-celled tissue, i.e. carcinoma sarcomatoides, and in addition there may even be areas of papilliferous columnar-celled tumour tissues. In the giant-celled carcinoma the multinucleate giant cells are only one item in the growth, the other



Fig 152. Papilliferous adenocarcinoma There is much fibrous tissue in this field (× 18)



Fig. 153. Metastasis of thyroid carcinoma in a lower cervical lymph gland from an impalpable primary in the thyroid. An example of so-called "Lateral Aberrant thyroid."

items being one or more of the other varieties of cancer tissue, especially spindle and polygonal celled carcinoma sarcomatoides.

The *papillary adenocarcinoma* (Fig. 152) is one of the commoner tumours and may sometimes be of relatively moderate malignancy. It tends to occur in younger people than do the others; it metastasizes commonly to lymph glands, rarely by the blood



FIG 154 Microscopic section of one lower cervical lymph gland shown in Fig 153 (×42)

stream. In size it is variable and its cysts and intracystic branching papillæ may be visible to the naked eye or only with the microscope. The epithelium is cubical or columnar; there may be differentiation into follicles. The so-called *lateral aberrant thyroid tumours* (Figs. 153, 154) belong to this group but behave in an odd chronic slow-growing way. At one time these swellings were regarded as papilliferous adenomata that arose in ectopic portions of thyroid tissue and sometimes became carcinomatous. Much ingenuity was displayed in tracing the embryological misplacement. It is now established that these tumours are secondary deposits in lymph glands from what is usually a tiny, slow-growing carcinoma in the gland proper. The latter may be quite difficult to find after the lobe has been removed, so minute may it be. But there is no doubt that the lateral tumours are metastases in lymph glands. The false theory of lateral aberrant tumours arose because in many of them all the lymphatic tissue had been replaced by tumour, but if enough of the tumours are examined with the microscope some normal lymph gland

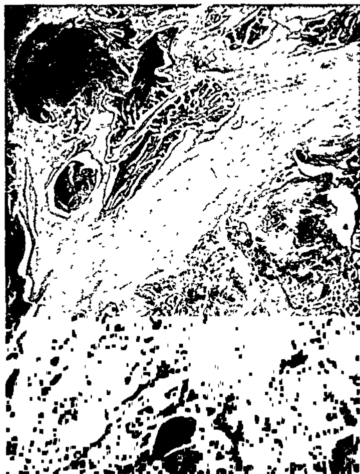


FIG. 152 Papilliferous adenocarcinoma There is much fibrous tissue in this field (× 18).



FIG 153 Metastasis of thyroid carcinoma in a lower cervical lymph gland from an impalpable primary in the thyroid An example of so-called "Lateral Aberrant thyroid"

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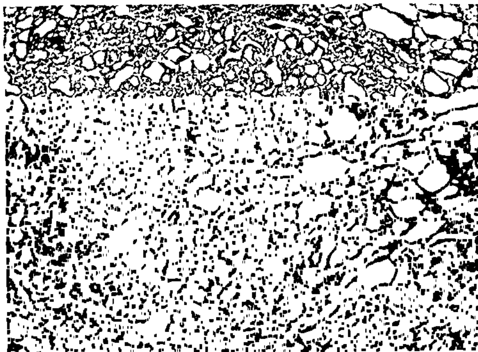


Fig. 155 Follicular adenocarcinoma Microscopic section of an encapsulated, slightly calcified growth (2 cm diameter) found in thyroid at necropsy four days after laminectomy for metastasis in a vertebra. There is no evidence of malignancy in this photograph nor in the rest of the section (×36).

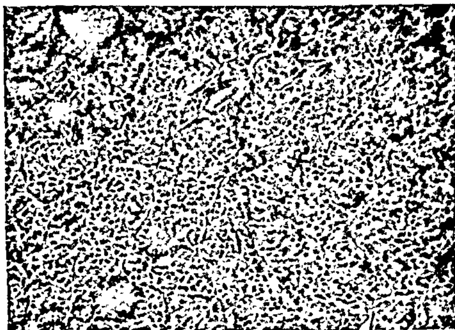


Fig. 156 Follicular adenocarcinoma Thyroidectomy for nodular goitre. Microscopic report not definitely malignant Patient died from metastases six years later Necropsy showed extensive secondary deposits in bones (×140)

tissue will be found. This type of carcinoma is rare; the masses occur in the cervical gland chain and may stretch down into the mediastinum: they may persist with little change over many years, even twenty or more. A really active malignant state may, however, supervene at any time.

The *follicular adenocarcinoma* (Fig. 155) may also vary greatly in size. Much of it is composed of acini containing colloid. It disseminates by both lymphatics and blood

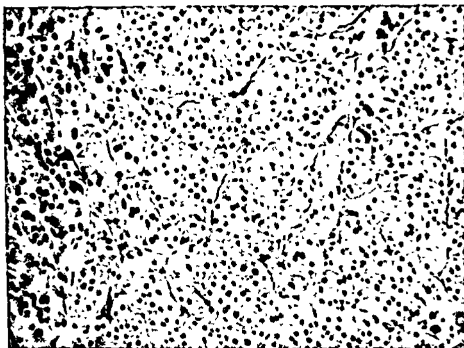


Fig 157. Hürthle-cell carcinoma Female 50 Microscopic section of one of several metastases in lower neck removed three years after thyroidectomy. The cells show a large amount of œsinophilic cytoplasm (× 150)

stream. Metastases by the latter route may rarely in part show almost normal thyroid tissue, which may function after the gland proper has been removed. This has led to the term "benign metastasizing goitre," a misnomer, since any tumour producing metastases must be malignant. A better term would be "carcinoma composed of almost normal thyroid tissue." Microscopically the follicular carcinoma shows in part differentiation into follicles, some with colloid, in part wide solid areas of cells. The *solid form of adenocarcinoma* (Fig. 156) is mostly or entirely composed of solid groups of polygonal cells. The groups are sometimes solid branching cords (the solid cord form of carcinoma). This type may resemble one form of nodular goitre and the differential diagnosis with the microscope may be difficult. It may in places have follicles containing colloid. The *Hürthle-celled carcinoma* (Fig. 157) is the malignant form of Hürthle-celled adenoma and is a rarity.

These forms of carcinoma are only moderately malignant compared to the small-celled anaplastic carcinoma which shows little or no differentiation into follicles, is highly invasive locally, and spreads readily and rapidly by both lymphatics and blood stream or to *squamous carcinoma*, which possibly originates from epithelial remnants of the

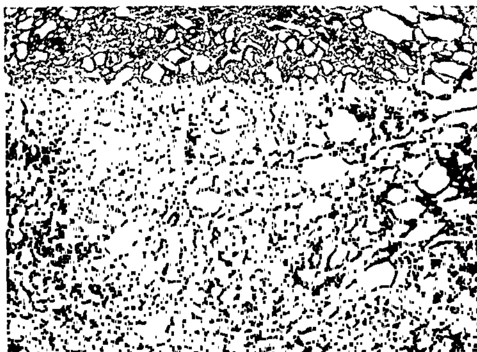


FIG. 155. Follicular adenocarcinoma. Microscopic section of an encapsulated, slightly calcified growth (2 cm diameter) found in thyroid at necropsy four days after laminectomy for metastasis in a vertebra. There is no evidence of malignancy in this photograph nor in the rest of the section (× 36)

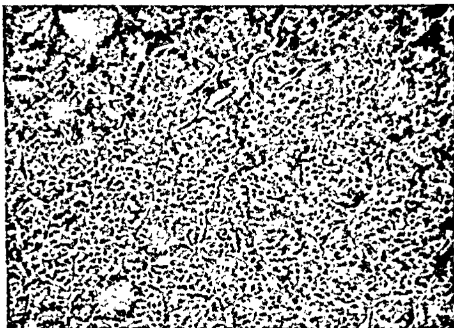


FIG 156 Follicular adenocarcinoma Thyroidectomy for nodular goitre Microscopic report not definitely malignant Patient died from metastases six years later. Necropsy showed extensive secondary deposits in bones (× 140)

experience in thyroid work diagnosed independently a subtotal thyroidectomy specimen as solid (parenchymatous) nodular goitre with no evidence of malignancy; the patient died 2 years later with widely spread metastases.

It may be difficult to distinguish between a papilliferous cystic adenoma and a papilliferous cystic carcinoma. The difficulty with some of the follicular lesions is notorious and gave rise to the phrase "benign metastasizing goitre," whereas the proper way to regard this phenomenon is that such a goitre is merely a carcinoma composed of tissue like that of a benign colloid goitre, thus breaking one of the rarely broken rules for carcinoma in general, that is, that carcinomata are seen with the microscope to differ greatly, or at least considerably, in structure from the normal tissues in which they arise. When there are many atypical cells in a goitre specimen the pathologist accepts this as indicative of carcinoma though it must be remembered that in the benign diffuse goitre of Graves' disease there may sometimes be moderately hyperchromatic nuclei much larger than the others.

Numerous mitoses are perhaps of greater value in the detection of malignancy in goitre than in other tissues.

In looking at a benign nodular goitre with the microscope the observer may see appearances simulating invasion of neighbouring normal thyroid tissue. It is generally accepted that carcinoma of the thyroid often invades veins; accordingly when the diagnosis is in doubt in a thyroidectomy specimen numerous sections ought to be taken to include long stretches of the periphery of the goitre so that as many peripheral vessels as possible will be seen But in a solid (non-follicular) nodular goitre a local area of hyperplasia is often seen to have compressed the wall of one of the wide very thin-walled vessels that are common in those goitres in such a way as to produce local invagination of the wall, and this may be mistaken for malignant invasion of the vessel.

It is likely that many benign goitres are dubbed carcinoma because pathologists do not recognize one or more of the pitfalls indicated above, and thus the number of cases of thyroid cancer alleged to be cured is falsely increased.

Spread. Direct spread to the adjacent structures leads to fixity to, and finally invasion of, the trachea, less commonly the œsophagus. Further spread afield in some types is almost limited to the lymphatic system and then the deep cervical glands are affected firstly. Spread by blood vessels occurs chiefly in the form of emboli but growth may extend in continuity along the thyroid veins into the great veins of the neck. The emboli settle most commonly in the lungs; other not uncommon sites are bones, brain and liver. Pulsating tumours of bone should arouse a suspicion of thyroid metastasis.

Symptoms, Signs and Diagnosis. Where a clear confident clinical diagnosis of malignant goitre can be made the condition has more often than not gone beyond the limits of satisfactory treatment; the prime diagnostic demand is the detection of features which arouse suspicion and lead to immediate further investigation and treatment. The high incidence of malignancy found in removed nodules by many writers has been adversely criticized (p. 308); there is greater justification for their strong advocacy of the removal of apparently benign nodules in the fact that the outlook in the established disease is bleak.

Suspicion of malignancy should be aroused when a visible and palpable lump in the thyroid is found to be hard and a history is given of a recent increase in its size. As the mass becomes larger and fixed and presses upon and invades surrounding structures

thyroglossal tract and behaves similarly to the small cell-type, or to *giant-celled carcinoma* (Fig. 158) which is probably the most malignant of all.

The *sarcomata* are rare. Some hold the opinion that the *reticulum-celled sarcoma* is a small-celled carcinoma.

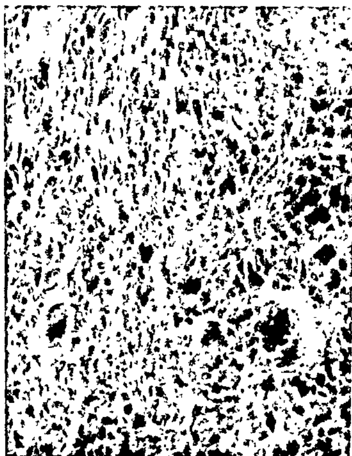


FIG. 158 Pleomorphic and giant-celled anaplastic carcinoma of thyroid gland Female 67 Patient died from metastases four months after thyroidectomy (× 150)

PITFALLS OF HISTOLOGICAL DIAGNOSIS

It is often extremely difficult to make a diagnosis with the aid of a microscope even when a subtotal thyroidectomy specimen is available. It is rash to attempt diagnosis on a drill biopsy specimen; a large piece of tissue is necessary. There are no specimens over which pathologists disagree more as to the presence or absence of malignancy than goitres. With the various forms of undifferentiated carcinoma and with squamous (epidermoid) carcinoma the diagnosis by microscopy is easy. But in any area of benign solid (non-follicular) nodular goitre where much fibrosis has occurred, the picture of cells in solid groups, with the groups varying in size and shape and scattered irregularly in abundant fibrous tissue, may resemble closely that of a scirrhous solid trabecular undifferentiated carcinoma, and deceive the pathologist. In the same type of goitre some of the minute nodules in the thyroid outside the larger nodules may be mistaken for carcinomatous invasion. The reverse mistake may less commonly occur; two histologists of considerable

Treatment. The methods employed in the treatment of malignant goitre may be resection, radiotherapy, radioactive iodine therapy, or a combination of these. The results on the whole are disappointing. The varied pathological types, however, suggest the probability of differences in prognosis and these are borne out by the published results.

A difficult and not uncommon problem arises when an apparently benign nodule is removed and is reported upon as being histologically malignant or possibly so. Nodular swellings, even if apparently benign on exploration, must be removed with a margin of normal gland and should not be simply shelled out. If they then are reported as malignant, any local break through the capsule will have been embraced, although invisible dissemination by lymphatic or blood vessel may have occurred. Should further operative procedures be undertaken? Provided the histological report has been made by observers highly skilled and experienced in thyroid pathology probably the correct courses are as follows. If the histologist regards the section as of doubtful malignancy but is unwilling to give a clear report of benignancy, no immediate further treatment should be given. But the patient should be re-examined at short intervals for a few years. Should the histologist be confident that the goitre is malignant, and preferably should that opinion be confirmed by other experienced histologists, then a more radical operation, as laid down below, should be advised. The alternative to this, should the patient demur, is radiotherapy or radioactive iodine therapy.

Where there is little or no doubt of the diagnosis either on clinical grounds or by findings on exploration, then radical surgery offers greater benefits, if the condition be operable, than other methods. Inoperability will be determined chiefly by strong fixation of the tumour to trachea or œsophagus, which fixity will probably indicate penetration, and by distant metastases. If the neoplastic mass be confined to one lobe, then that lobe, with the isthmus and possibly part of the other lobe, should be totally resected, and a block dissection of that side of the neck performed, whether or not there are enlarged glands. The block dissection, of course, also entails removal of the sternomastoid muscle and the internal jugular vein.

Where the neoplastic process has invaded both lobes a total thyroidectomy is needed. This will be accompanied by a block dissection of glands as above, on the side most indicated; on the other side a similar dissection will be done but the internal jugular vein will be preserved; all veins, however, entering it from the thyroid must be divided close to it to deal with any permeation as thoroughly as possible.

Total thyroidectomy and total lobectomy naturally entail great care in the preservation of laryngeal nerves and parathyroid gl[illegible] deep irradiation of the appropriate areas. [illegible] best results are obtained by surgery, even if [illegible] X-ray treatment.

The treatment of that curious type of papillary adenocarcinoma which used to be designated *lateral aberrant thyroid tumours* (p. 311) needs particular comment. While the condition may remain stationary, or be only slowly progressive for many years, its change towards more rapid spread is sufficiently frequent as to demand removal of the masses together with the lobe of the gland on the affected side. It is doubtful, however, if a block dissection, with all that that entails, is necessary. Thereafter a close supervision of the patient should be maintained.

there occur dyspnœa, dysphagia, alteration in the voice and perhaps stridor from recurrent nerve palsy, pain in the neck, shoulder and arm from involvement of the superficial sensory nerves, Horner's syndrome from pressure upon the cervical sympathetic chain, and bradycardia from implication of the vagus. Loss of weight is a common feature. Unequivocal associated hyperthyroidism is an extremely rare phenomenon.

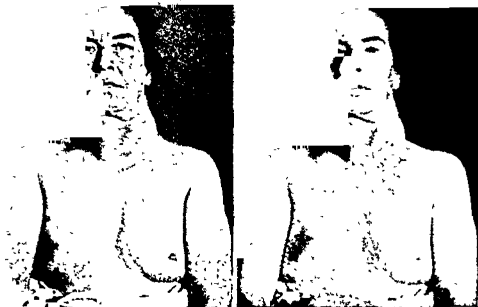


FIG 159 Carcinoma of thyroid. Well-marked clinical case of carcinoma of the thyroid in female age 56 Note the dilated veins in chest wall in the infra-red photograph

Metastases, in lymphatic glands or elsewhere, may be the first presenting features; the parent tumour in the thyroid may then be small and even impalpable. Radiological examination of the chest is an imperative proceeding since there may be more evidence of the nature of the disease in the chest than in the neck.

The difficulty of early diagnosis has added to the advocacy, on more doubtful grounds (p. 308), of the removal of all nodules, particularly apparently single ones. A rather hard swelling, smooth or irregular, steadily increasing in size, is to be regarded with suspicion. The hardest swellings in the thyroid are due to hyaline fibrosis or calcification of nodules, to carcinoma, or to Riedel's thyroiditis. The last may affect only one lobe at the time of examination and simulate a localized swelling. Again, calcification can be associated with slowly growing neoplasms. Hashimoto's disease may be mistaken for carcinoma, but the gland is firm rather than hard, is usually symmetrically enlarged, and has generally reached a size where the absence of glandular metastases is noteworthy. Sudden or very rapid increase in thyroid swellings is more likely due to hæmorrhage or to inflammation. Rare cases of syphilis of the thyroid have been described in which there was great simulation of neoplasm even to infiltration of the trachea. The thyroid may also be invaded by a squamous carcinoma of the pharynx or upper œsophagus; a history of the early onset of dysphagia should arouse suspicion of this possibility.

irradiation. Since these highly malignant tumours spread quickly to the lungs the irradiation is aimed at both neck and thorax. Should the neck tumour not recede, however, with this technique, the attack is massed upon it. While the immediate results in some patients may be almost dramatic, the tumour melting away in a few weeks, the end results are not good, chiefly because of distant spread. But even where less striking recession takes place much local relief may be obtained from the pressure exerted on the trachea by large tumours. Because of this, tracheotomy should be avoided, as also because disturbance of the tumour in its performance may lead to dissemination.

The advent of *radioactive isotopes* of iodine opened up a new, if limited, method of attack. If the tumour will, or can be made to, take up the radioactive iodine, the malignant cells can be destroyed; this effect is largely due to the Beta radiation. Most malignant thyroid tissue, however, picks up little or none of the radioactive iodine since most of the tumours show little differentiation to follicular structure. The small-celled and the papillary, solid-cord and Hürthle-celled adenocarcinomata present little or no differentiation. On the other hand, although in a minority, there are some adenocarcinomata, particularly of the follicular type, which will pick up the radioactive iodine and their cells will be irradiated, as of course will those of their similarly constructed metastases. The radiation effect from the colloid spreads only for about 1 millimetre and thus other tissues are not irradiated.

The pick-up of radioactive iodine can be greatly increased in such differentiated tumours and their metastases by the removal or the destruction of the normal thyroid tissue. The latter, thyroid ablation, is achieved either by total thyroidectomy, or by destruction of functional normal tissue by radioiodine, or by thiouracil, which blocks the uptake of iodine by the thyroid. Whether it is that the normal thyroid competes too strongly for the iodine against the tumour and its metastases, so that little is absorbed by the last two while the first is still present, or whether it is that the tumour or its metastases increase in functional activity to take over the role of the ablated thyroid, is still a debatable point.

From the practical point of view it is important that an idea of the degree of differentiation of the tumour [illegible]
isotopes is worthw [illegible]
of tumour as possible. If differentiation be present in any degree the next step is to ablate the normal thyroid tissue, preferably by total thyroidectomy, if not, by a destructive dose of radioiodine. Thereafter, if a test dose of radioactive iodine be given and it is found, by means of the Geiger counter, that the tumour or its metastases are concentrating the iodine, then successive therapeutic doses of radioactive iodine are given at intervals of 2–4 months until the Geiger counter cannot reveal any further point in the body where iodine is being concentrated. Myxœdema naturally will develop and have to be controlled. The myxœdema may wax and wane until it is finally totally established, since metastases will take on the functions of the ablated normal thyroid tissue until they are destroyed.

Such treatment by radioactive isotopes of iodine have resulted, in the appropriately differentiated tumours, in apparent cure in some instances and in the amelioration of the condition in others, with a prolongation of life for several years.

Prognosis. The outlook depends chiefly upon the histological type of the tumour, and rather less upon the stage it has reached when advice is sought. Long-term follow-up statistics are hardly to be found and this suggests that the ultimate results are poor

Irradiation Treatment. Irradiation may be carried out by deep X-ray therapy or by the use of radioactive isotopes of iodine. External irradiation by applicators of radium has proved ineffectual. Deep X-ray therapy is best combined with surgical excision where



FIG 160 X-ray of chest in a patient with primary carcinoma of the thyroid The goitre extended retrosternally and involved the left recurrent laryngeal and left phrenic nerves

the latter is practicable; too often it is not. The effects of radioactive iodine are dependent upon the ability of the tumour to take up the isotope This is a matter of some variability.

Deep X-ray Treatment has proved of most value in the least differentiated tumours, although more than half of these are insensitive to radiation; the differentiated tumours are markedly insensitive. While the biopsy may show the lack of differentiation it is not possible to predict from its histological picture that a particular tumour will react to

Symptoms and Signs. The great majority of cystic hygromata in the neck cause no symptoms since most are mainly superficial and the skin of the neck bulges out with their growth to obviate any deep compression. In a few, however, the deep cysts cause



FIG. 161. Cystic hygroma bulging into the floor of the mouth and interfering with swallowing.



FIG. 162. Large cystic hygroma in child age 4.



FIG. 163. Cystic hygroma excised from axilla.

dyspnœa, even to stridor, by tracheal compression and shift or, where they have passed into the mouth (Fig. 161) interfere with the passage of food.

The swellings vary in appearance and size. Some are obviously multiloculated (Fig. 163), some present a smooth ovoid appearance. Usually they are tense to the feel. All are highly transilluminable unless hæmorrhage has occurred. Some exhibit no change in size over years, some grow slowly, some get bigger suddenly without the aid of hæmorrhage, very rarely one may rapidly become smaller. Infection is uncommon but is a serious event because of its potential spread throughout the neck.

although it is to be remembered that the average age of patients contracting the disease is about 55 years. The papillary adenocarcinoma is the most benign, the giant-celled carcinoma the most malignant. The operability rate lies between 50 and 60 per cent; it is about twice as great where the carcinoma has supervened on a pre-existing goitre as where it has occurred in an apparently normal gland. Overall 5 year survival rates in published series approximate to 50 per cent after surgical excision, 60 per cent after excision and radiotherapy. The 5 year survival rates are given as 50–80 per cent in papillary carcinoma, 30–40 per cent in follicular carcinoma and 0–15 per cent in giant-celled carcinoma. But it is difficult to determine from publications the prognoses of the different types since authors differ so much in their classifications. The writer believes that the survival rates given above are too high because of the inclusion of benign cases faultily diagnosed as malignant by pathologists inexperienced in thyroid histology

Cystic Hygroma

A cystic hygroma is a unilocular or multilocular swelling which usually contains thin clear fluid within a fibrous, endothelial-lined wall. It is by far most commonly found in the neck; other sites are the axilla, chest wall, groin and thigh. It is sometimes called cavernous lymphangioma but its sites of origin are generally different from those of ordinary lymphangiomas in which the individual cysts, although the mass containing them may be fairly large, are always minute, as against the spaces of up to several inches diameter of cystic hygroma. While the main bulk of a cystic hygroma is usually superficial, outlying processes commonly pass extensively through and between adjacent and subjacent structures, particularly muscles.

The *ætiology and pathology* of the condition are still not clear. It is generally believed that they are derived from lymphatic sacs known to exist in the human fœtus; there is controversy as to whether these sacs are derived from the jugular or other veins or whether they develop independently from mesenchyme and later have connections with the veins. One theory is that solid cords of endothelial cells grow out in many directions from sequestrated lymphatic tissue, passing between muscle fibres, nerves, vessels and other structures. In time these cords hollow out irregularly and become more or less distended with fluid, as permitted by their anatomical situation, so that large sacs may have smaller ones communicating with them or there may be many independent sacs. The tissues through which the cords originally passed may thus become separated from one another and covered with endothelium; this explains why a structure such as a vessel, nerve or muscle bundle may at times be found crossing the cavity of a cyst.

The cyst wall is white or grey, often quite tough, and apparently almost avascular. Hæmorrhages, however, occur sometimes into these cysts and may be the reason for the detection of a swelling hitherto unnoticed.

In a small proportion of cases there is an associated lymphangiomatous condition of the tongue.

The cysts are most commonly obvious at birth; the great majority are obvious by the end of the second year. There is no relationship of size to age, nor is there any difference of incidence in the sexes.

It is sometimes said that there is a great mortality rate from this lesion; it is difficult to know on what this is founded.

Embryology

There is disagreement on the origin of these anomalies. The original conception, the branchial theory, was that they were derived from failure of obliteration of the embryological precervical sinus. The latter is formed by fusion of the second and fifth branchial arches and is normally a transitory sinus which has disappeared by the fifth week of fœtal life. The complete fistula opens into both neck and pharynx but the orifice into the latter cannot occur developmentally and must be regarded as a secondary breakdown if one accepts the branchial origin. Partial persistence at either end leads to a sinus into the neck or into the pharynx; persistence of a central portion leads to a branchial cyst.

The branchial theory is strengthened inasmuch as small fatty excrescences containing cartilage are occasionally found in the same position in the neck as the opening of a



Fig. 164 Bilateral branchial fistulæ. Mother and daughter. Note the cystic collection on the right side in the child.

fistula, thus, it is claimed, constituting homologues of the pinna which is developed in the first branchial groove. These excrescences do not usually have an associated opening and their removal is a simple superficial operation. Some believe the origins of the fistula to be entirely concerned with the second branchial cleft, because the fistulous tract passes between the two carotid arteries.

The branchial theory [illegible]

emb [illegible]

can be present below the level of the hyoid bone. The track is then attributed by some of them to the persistence of the thymic duct.

Branchial Fistula. A branchial fistula is present from birth and its presence is more often made known in infancy by the repeated appearance of a tiny drop of clear fluid at the same spot in the neck rather than by any obvious orifice. For the external opening is always small and may be as minute as to cause difficulty of detection. With the passage of the years the opening tends to become more obvious and this is especially so when attacks of infection have supervened; a small dusky red areola then surrounds the opening and the skin above the orifice presents a small crescentic fold, just as a persistent thyroglossal sinus does, with the concavity downwards.

The opening is always in line with or very near, the anterior edge of the sternomastoid muscle and commonly about the level of the junction of the lower third and upper

X-ray may show extensions into the thorax and dislocations of larynx and trachea in some of the largest tumours.

Treatment. It was at one time held that spontaneous disappearance of the tumour would occur if it were ignored long enough; this was attributed to the effects of repeated mild attacks of inflammation. While this view is not now in favour it cannot be denied that it is extremely rarely that one finds a cystic hygroma in an adult, even a young adult. It might be expected that operation or other treatment would occasionally be refused, or skilled treatment be unavailable, or inadequate operations be performed often enough (since complete extirpation is sometimes impossible) for such tumours to be seen occasionally in adults. Yet they are not. There may therefore be some tendency towards spontaneous obliteration.

Some form of active treatment is however indicated for the majority of cases since the tumours are unsightly, since they may become infected especially after upper respiratory infections, and because in a few instances the pressure features require relief.

The most effective forms of treatment are surgical excision and the injection of sclerosants. Repeated aspiration usually ends in sepsis and aspiration should be reserved to relieve serious pressure temporarily. Deep X-ray therapy has not proved of value.

Sclerosants sound theoretically ideal. But some of the large tumours have a multitude of small complete deep cysts which would require very many injections, apart from the difficulty of finding them all with the point of the needle. Another objection to sclerosants is that occasionally there seems to be a communication between the hygroma and the venous system, as presumptively shown by the quick disappearance of injected lipiodol in one authentic case. None the less, sclerosants are employed by some surgeons and, if used cautiously and repeatedly, may achieve the purpose.

The main line of treatment, however, is surgical extirpation. Generally, if the operator has patience he can find his way easily by blunt dissection around the outer wall of the great mass of the tumour without opening it or at the most, opening into a few of the smaller cysts. It is when the deep processes are reached that difficulty is encountered. Main vessels, nerves and other structures have to be carefully defined and preserved. Usually the operator will feel dissatisfied with the deep dissection, knowing that he must have left little pockets between muscle strands and so on. But if he has been as thorough as seems reasonable he will not often have recurrences, these tiny residual pockets seem usually either to atrophy or remain as they were.

Branchial Cysts and Fistulæ

Because of doubts as to their origins the terms *lateral cervical cyst* and *lateral cervical fistula* are probably better names for these entities but time has somewhat hallowed the retention of the old titles. The "fistula," however, is sometimes only a sinus and the terms *branchial sinus* or *fistula* and *cervical sinus* or *fistula* are used synonymously; it is usually impossible on clinical examination to determine whether the condition is a sinus or a fistula.

The cyst forms a smooth somewhat globular swelling of variable size in the upper lateral aspect of the neck. The fistula is clinically evident as a tiny orifice in the skin in line with the anterior border of the sternomastoid muscle somewhere in its lower half; the orifice leads into a much wider tubular track of variable length which may reach as high as and communicate with the pharynx in the supratonsillar fossa.

and the patient or the parents seek advice. A more acute infection occasionally erupts and there then occurs a tender swelling along the line of the deep track.

Because of the small opening it is by no means always possible to pass a probe along the track. Such probing has, as has also occurred with injections of lipiodol, sometimes led to features suggestive of vagal irritation such as palpitation, an irregular pulse, and cough. The vagus nerve is certainly a close relation in the higher part of the course of the track.

The track itself is usually a quite well formed cylindrical structure, which can be expanded with injected fluids to a diameter much greater than that of the external orifice. A complete fistula from neck to pharynx is not uncommon since patients who have had fluids injected through the skin orifice quite occasionally assert that they can taste the fluid although they fail to spit it out in quantity; this confirms that the pharyngeal opening is minute in size. Most "fistulæ" are, however, incomplete and are therefore really sinuses. Blind openings into the supratonsillar fossa have been found at autopsies, but these do not seem to have caused trouble during life.

Treatment. All forms of treatment other than operative removal of the track have proved entirely unsatisfactory. It is sometimes stated that a two-fold approach, from both neck and pharynx, is necessary. But dissection from the neck only, reaching as high as possible towards the pharynx, has not been followed by any recurrence in the writer's series. The essentials of adequate removal are that a wide exposure is made in a bloodless field and that a probe is kept in the track either as high as the track exists or up to the point where the track turns sharply deeply under the posterior belly of the digastric muscle and between the internal and external carotid arteries. The bloodless field is achieved by the subplatysmal infiltration of saline-adrenaline solution of 1 in 150,000 strength (1 ml. of 1000 adrenaline added to 150 ml. normal saline); the adequate exposure is attained by reflection upwards of a large flap. To obtain the latter a long transverse incision, with a central ellipse containing the opening of the fistula, is made in the line of a skin crease. If this incision is long enough there is rarely need to make the two parallel incisions at different levels which are sometimes advised. A neat long single incision is less noticeable than two short ones. A long almost vertical incision over the track should never be made. A probe within the track is helpful and if it cannot be got through the skin opening it can be introduced without difficulty after the skin incision is made. Lipiodol and methylene blue have been used to help define the track at operation but the writer has found them unnecessary. It is advised by their advocates that the medium is introduced and a purse string suture then tied round the orifice.

Each flap consists of skin, subcutaneous tissue and platysma. The lower flap is raised only half an inch or so to make accurate suturing easy at the finish. The upper flap is dissected as far as the length of the incision will allow. Gentle traction on the ellipse of skin and use of the indwelling probe will show up the line of the track in the muscular fibres of the anterior border of the sternomastoid muscle. These fibres should be taken with the contained track; the latter alone is too tenuous a structure by itself to withstand much manipulation. The track is easily dissected as far as the posterior belly of the digastric muscle. Its further course is best followed using blunt dissection with a small curved Mayo's scissors. Firm traction will now more easily cause rupture and must be avoided. The track, however, usually peters out and breaks as one passes between the carotids. As has been said, recurrence does not follow if one has got to this point. There

two-thirds of the muscle; at birth it may be somewhat lower, in adult life higher, than this. Rarely a deep cordlike thickening can be felt running upwards from the opening.

Occasionally instead of the discharging skin puncture there develops slowly in this area a flabby cystic swelling surmounted by a white dot representing the usual point of



FIG. 165 Branchial fistula. Girl age 6½.



(By courtesy of the Editors of the "Practitioner")

FIG. 166. Lipiodol injection of branchial fistula

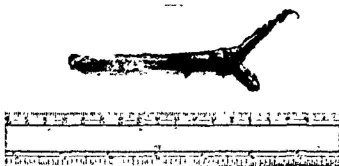


FIG 167 Branchial fistula Excised specimen.

opening. In time this point gives way and there is a discharge of clear fluid; the opening in this type may close from time to time.

The track is lined by squamous or columnar epithelium. There has been no sex difference in the author's series of some forty cases. The condition is occasionally bilateral. The author has on two occasions observed mother and child with the condition.

The fistulæ may go on discharging clear thin fluid and, if the amount be small, the patient may do nothing about it and not seek treatment even in adult life. More frequently, however, a mild infection supervenes. This produces a semi-purulent discharge

is no need to attempt any ligature of the upper end. The platysma and skin are then approximated without drainage.

Branchial Cysts

A branchial cyst (lateral cervical cyst) forms a smooth swelling, somewhat globular or ovoid in shape, bulging outwards in the upper lateral aspect of the neck. Part of the swelling lies deep to the sternomastoid muscle; the main mass projects forwards and



FIG 170 Lower cervical dermoid cyst Female 23 Swelling noticed since a baby. Photograph on right shows the excised cyst measured in inches.

slightly downwards in front of it. It may in size develop to the bulk of a large orange. Its most characteristic feature, although not an unexceptional one, is its relative flaccidity. There is an absence of tension to the feel which gives an impression that the cyst wall could easily accommodate more fluid. This is an important characteristic since the chief diagnostic differentiation is from a tuberculous abscess of lymphatic glands: the latter never gives any impression of flaccidity whereas a branchial cyst can intermittently swell somewhat and become tense; this is probably the result of a temporary increase in the fluid content from some mild infection. The cyst contains usually the characteristically shaped crystals of cholesterin, and some writers advocate aspiration of the cyst and examination of the deposit under the microscope for what they regard as conclusive evidence.

While a branchial fistula is always present at birth a branchial cyst rarely makes itself obvious in the first decade of life and is a feature more of the second and third decades. Its appearance commonly follows some upper respiratory infection, just as does a thyroglossal cyst or fistula. Such a sequence of events suggests, it has been argued, that possibly the branchial remnant was draining into the pharynx until the infection caused a permanent closure of the pharyngeal communication. But it would seem more likely, if one considers the process as parallel to thyroglossal tract affairs, in the great majority of which there is no upper communication, that the gradual increase of fluid is due to a mild infection in the wall.

Whatever be the cause the cyst increases slowly in size and quite frequently ceases



FIG. 168. Right branchial cyst. Female 21. Painless swelling for 2 years.



FIG 169. Sublingual dermoid cyst. Female 19 Painless swelling since childhood.

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FIG. 169. Sublingual dermoid cyst Female 19 Painless swelling since childhood.

In the great majority of cases the swelling rapidly subsides, but occasionally suppuration occurs. Fluctuation must not be expected if abscess formation occurs deep in the neck.

If antibiotics are administered suppuration may be masked and tenderness may be very slight.

Acute retropharyngeal adenitis only occurs in infants and will give rise to dysphagia. A bulge will be seen or felt in the posterior pharyngeal wall which will be fluctuant when abscess formation occurs.

Diagnosis. The diagnosis is usually obvious and is characterized by the presence of an acute primary lesion, the rapid onset, fever and the hot brawny tender enlarged nodes. Confusion may occur in differentiating acute adenitis from an infected branchial cyst or an infected cystic hygroma but the past history will be of great help.

Occasionally submaxillary sialoadenitis may be mistaken for submaxillary adenitis, but examination of the floor of the mouth will reveal the presence of a stone in the submaxillary duct.

Careful examination of the ear will clear up any difficulty in distinguishing acute mastoiditis from acute inflammation of the mastoid lymph node.

Difficulty may also occur in differentiating acute cervical adenitis from the acute type of tuberculous cervical adenitis and in some cases acute infection may occur in chronic tuberculous glands.

The differential diagnosis of neck swellings generally is discussed on page 333 (under Tuberculous Cervical Adenitis).

Treatment. In addition to treatment for the primary infective lesion, treatment will be required for the acute cervical adenitis.

General treatment will consist of rest in bed if the constitutional disturbance is marked and administration of a suitable antibiotic such as procaine penicillin 300,000 units once daily. Locally hot dressings such as kaolin poultice should be applied.

If suppuration occurs an approach through an incision in line with a skin crease is indicated. An incision made too early may spread the infection whilst too great delay may result in the pus burrowing in the neck and even into the pharynx and mediastinum. The abscess should be opened by Hilton's method and curettage and all unnecessary trauma should be avoided. Corrugated rubber drains should be used rather than tubes and need not be retained for longer than three days. Dressings should be done only once daily and fomentations are unnecessary after drainage has been performed.

Acute retropharyngeal abscess should be opened into the pharynx through the mouth with a sinus forceps.

Chronic Non-Specific Cervical Adenitis. The majority of cases of chronic apparently simple cervical adenitis are in reality due to tuberculosis. However, there are cases in which it is impossible to demonstrate the presence of tubercle bacilli and on culture streptococci or staphylococci will be found.

Usually it follows an attack of acute cervical adenitis and is then due to persistence of the primary infective lesion, e.g. carious teeth or a chronic or subacute infection of the scalp.

Clinically the glands are only moderately enlarged, painless, discrete and elastic. Suppuration is rare and when it occurs tuberculosis should be strongly suspected. Treatment is directed towards removal of the cause.

growing when it has got to about a diameter of three inches. The swelling is painless unless infection supervene; such infection would seem to come usually from the pharynx or to be blood-borne. Suppuration may occur but attacks of milder infection are more common and these resolve spontaneously. When suppuration occurs, unless an incision be made, the cyst, it is said, may rupture spontaneously on the surface of the neck or, very rarely, into the pharynx. The author has not seen these events; any infective process seems mild and tends to die out spontaneously. No treatment other than complete removal by dissection is of value. If, after exposure of its superficial surface the capsule is adhered to, little difficulty is encountered, particularly if it is remembered that the flattened internal jugular vein is a close relation of the swelling. A deep projection from the cyst towards the pharyngeal wall is occasionally met with.

Acute Cervical Lymphadenitis

The cervical glands are commonly the seat of acute inflammation. The infecting organisms, usually hæmolytic streptococci or staphylococci, reach them by lymphatic channels from various sources the chief of which are the mouth, fauces, face, and scalp. The site of the primary infective lesion determines the lymph node involvement and this is usually the upper deep cervical group or the submaxillary group. Acute cervical adenitis may also be seen in association with infectious diseases such as diphtheria and scarlet fever. A special type of acute cervical adenitis occurs in Cat Scratch Disease which is due to a virus infection related to lymphogranuloma.

Pathology

Although hæmolytic streptococci and staphylococci are the usual organisms, others such as B. mallei, B. pestis and B. tularensis are occasional causes. The glands become enlarged, hyperplastic and show increased vascularity. At first and in moderate infections the glands are discrete but in the severer infections the surrounding connective tissues are involved and the glands become matted together. In the severest infections the process spreads into the whole deep chain of glands and the cellular tissues of the neck. Microscopically the glands show proliferation of the lining endothelial cells and infiltration with polymorphs. The inflammatory process usually resolves completely, but may proceed to suppuration, particularly in children. The resulting abscess may track in the neck or discharge through the overlying skin.

Clinical Features. The symptoms will depend on the type and virulence of the causative organism. The portal of entry of the organism is usually readily seen and this primary infective lesion may be the only complaint. Occasionally it may be quite insignificant, particularly in streptococcal infections and in tularæmia.

The onset is usually fairly rapid and the constitutional disturbance varies from a feeling of malaise and slight pyrexia to high fever, delirium and even collapse. The patient will complain of a throbbing painful swelling in the neck but the pain may be referred down to the shoulder or up to the ear. At first the glands will be tender, enlarged and discrete. Later they will give rise to a hot, tender brawny swelling and the overlying skin may be red. In severe instances the whole side of the neck may be swollen and hard and tender with a combination of lymphadenitis and cellulitis; the constitutional features are marked; this form is now rarely seen because of the introduction of chemotherapy and antibiotics. There may also be some degree of torticollis, particularly in c

It is convenient to refer to the three types as upper cervical, lower cervical and diffuse tuberculous lymphadenitis.

Pathology. Both human and bovine strains of tubercle bacilli have been found in tuberculous cervical adenitis and their relative frequency has been a matter of research and argument. Infection by bovine bacilli is naturally directly related to the consumption of infected milk and is therefore more common in places where milk is neither tuberculin-tested nor pasteurized. With increased safeguards in public milk supplies the relative



Fig. 173 Tuberculous cervical adenitis Lower cervical type



Fig 174 Tuberculous cervical adenitis Caseating mass of glands removed at operation.

frequency of bovine infection has greatly diminished. The incidence in Scotland has always been regarded as higher than in England, as indeed it is also in rural districts where the consumption of milk tends naturally to be unregulated. An interesting trend in the change of incidence is that, whereas previously children under 5 were most commonly affected, the higher figure is now far above that age; this seems attributable to the more widespread use of dried milks in infancy and babyhood, and to the increased consumption of milk after this age under schemes for its provision in schools.

The response of lymphatic glands infected with tubercle bacilli is an allergic one and not due to the production of toxins. There is an exudative and cellular reaction; the glands swell, and a diffuse sticky periadenitis binds them quickly to one another and to adjacent structures. Within the glands tubercles are formed. The further progress of the disease depends upon the balance of the virulence of the infection and the resistance of the patient. Where these are most favourable the periadenitis resolves and leaves the individual glands more palpable and mobile; in time there is spontaneous disappearance of the enlargements. Under rather less favourable conditions, caseation takes place at points in the mass and (Fig. 174), while the periadenitis disappears and perhaps discrete

Tuberculous Cervical Adenitis

Ætiology. Tuberculous cervical adenitis, though much diminished in incidence and severity in the past thirty or forty years, is still not an uncommon disease. Its occurrence is related chiefly to the ingestion of infected unpasteurized milk and to bad social conditions such as overcrowding, whereby infected sputum is swallowed or inhaled. There are three main types of the disease. In the first of these, by far the commonest type, the upper deep cervical glands are the earliest to be involved and the disease more often than



FIG 171 Tuberculous cervical adenitis The initial, and often the only site, in the upper cervical type.



FIG. 172. Tuberculous cervical adenitis Upper type showing involvement of the whole cervical chain with multiple sinuses

not remains restricted to these (Fig. 171); further spread, if it occurs, is downwards and forwards to involve the other glands of the deep cervical chain (Fig. 172). The route of infection in this type is usually through the tonsils but may be elsewhere in the nasopharynx or pharynx, rarely in the middle ear or on the face. The condition is then, in the terminology of tuberculosis, a "primary complex," signifying the combination of primary focus and associated lymphadenitis. This type is seen chiefly in childhood and young adult life.

The second type, most common in adults, affects the lower cervical glands (Fig. 173) and is an extension of the infection upwards from tuberculous mediastinal glands, the primary infection being from the lung hilum into the hilar glands.

In the third type there is a widespread enlargement of all the cervical glands. These remain rubbery and discrete, often simulating lymphadenoma, and do not go on to caseation as happens in the other types. The infection is believed to reach them by the blood stream from some primary focus elsewhere.

be left after almost complete resolution; a mass may remain indolent, with little or no change in size, for many months or even years. If caseation and softening occur at various points, there may be a coalescence to form an abscess which then perforates the deep fascia in front of the sternomastoid, or the sternomastoid itself, to become subcutaneous. The condition is now that of a deep partly solid, partly liquefied mass and of a subcutaneous collection of pus, the two connected by a narrow opening, the whole then known as a "collar-stud abscess." The skin overlying this swelling then slowly reddens at one point (Fig. 175), maybe at a distance from and not overlying the deep opening, thins and finally ruptures to allow the egress of the pus and the formation of a sinus, through which secondary infection will enter. Such abscesses may form and so behave at many points on the neck (Fig. 172). The skin surrounding the sinuses may become heavily infected and necrotic.

The constitutional features associated with the disease are rarely severe. Even where the onset is acute there is usually only moderate malaise and the temperature is but slightly raised; the patient, usually a child, resents being restricted of movement. In more seriously affected patients there is, however, languor and loss of appetite.

In *lower cervical tuberculous adenitis*, commonest in early adult life, there is swelling of the glands deep to the lower end of the sternomastoid and further out in the posterior triangle. The tendency is for slow caseation and abscess formation to occur and, if proper treatment be not afforded, for the formation of multiple sinuses (Fig. 176). The constitutional features are again variable; in the main they are not severe. X-rays will usually show the accompanying involvement of mediastinal glands.

In *diffuse tuberculous cervical adenitis*, the relatively rare form of the disease, the glands tend to be rubbery and discrete, thus simulating those of Hodgkin's disease. Any tendency to caseation and softening is slow and relatively uncommon. The constitutional features are of minimal degree. A biopsy is commonly required to establish the diagnosis.

Differential Diagnosis. The differential diagnosis of tuberculous cervical adenitis brings in nearly the whole range of swellings in the neck. Swellings of the thyroid, however, are unlikely to be mis-diagnosed. Swellings of the parotid and submaxillary salivary glands should also be readily recognizable by their limitation to their anatomical location. The chief difficulties arise in differentiation from other forms of enlargement of lymphatic glands, such as are caused by non-specific infections, by glandular fever, by the reticuloses and by secondary malignant neoplastic disease, and in differentiation from such other conditions as branchial cyst, carotid body tumour, and cervico-facial actinomycosis.

Non-specific pyogenic glandular infections may be acute or chronic. In the acute form, which may be secondary to infections of the throat, face or scalp, there is a rapid tender swelling of the glands and of the tissues surrounding; the overlying skin becomes red; a brawny cellulitis of the whole side of the neck may develop. Constitutional symptoms may be marked. It is often impossible to differentiate the less severe instances of this type from the acute type of tuberculous adenitis. Failure of simple chemotherapeutic measures and persistence of the swelling for a month should lead to the likely diagnosis of tubercular infection. Chronic non-specific infection is uncommon and occurs chiefly secondary to persistent infected areas on the scalp; the glands are usually small and shotty and have little tendency to suppuration.

swellings result from the mass, the swellings remain for a considerable period before receding. Radiographs taken years later will show that these caseous areas have become calcified. Occasionally the caseous areas coalesce to form one large cold abscess which does not track to the surface but develops a tough wall over a period of years. In still less favourable instances the caseous pus breaks out beyond the bounds of the mass and tracks to the surface. In the least favourable cases the infection spreads to adjacent groups



FIG 175 Tuberculous cervical adenitis, upper type, showing subcutaneous abscess with thinning and infection of the overlying skin.



FIG. 176 Tuberculous cervical adenitis, lower type, showing multiple sinuses.

of glands in the line of the lymph drainage and eruption of cold abscesses to the surface of the neck takes place at many points.

Clinical Course. In the common *upper cervical type* there appears, gradually or suddenly, a swelling or swellings just below and behind the angle of the jaw. Most commonly the swellings remain limited to this area but where infection is severe and resistance low then more swellings appear further downwards and forwards in the neck (Fig. 172). Where the onset is slow and insidious it is usually possible to palpate several individual swellings; where the onset is acute there may be apparently only one palpable large swelling due to the coalescence of several affected glands. A lesser degree of swelling of the glands on the other side of the neck is a common concomitant.

Where the swelling is rapid and accompanied by severe or moderately severe constitutional symptoms it will not be possible to differentiate the condition from an acute non-specific infection, usually streptococcal, until failure to respond to chemotherapy and persistence of the swelling for a month or so make the diagnosis clear.

The further clinical course of the disease follows upon the possible pathological changes enumerated above. Complete resolution may occur; small shotty residues may

be left after almost complete resolution; a mass may remain indolent, with little or no change in size, for many months or even years. If caseation and softening occur at various points, there may be a coalescence to form an abscess which then perforates the deep fascia in front of the sternomastoid, or the sternomastoid itself, to become subcutaneous. The condition is now that of a deep partly solid, partly liquefied mass and of a subcutaneous collection of pus, the two connected by a narrow opening, the whole then known as a "collar-stud abscess." The skin overlying this swelling then slowly reddens at one point (Fig. 175), maybe at a distance from and not overlying the deep opening, thins and finally ruptures to allow the egress of the pus and the formation of a sinus, through which secondary infection will enter. Such abscesses may form and so behave at many points on the neck (Fig. 172). The skin surrounding the sinuses may become heavily infected and necrotic.

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In *glandular fever* the swellings may be found elsewhere than in the neck, although sometimes they may be apparently confined to it. They are diffuse, discrete, tender and tense. Usually moderately enlarged, they may on occasion become as large as a plum; *there is no tendency to suppuration. There is a mild intermittent fever, the blood shows* a lymphocytosis and the diagnosis is established by the Paul Bunnell test. In *lymphadenoma* (Hodgkin's disease) the glandular swellings are of moderate size, soft to firm, smooth, discrete, and not tending to caseation; they simulate closely the diffuse form of tuberculous adenitis. Glands elsewhere are also commonly affected; the spleen is palpable in about one-third of cases. Intermittent fever (Pel-Ebstein) may occur. Biopsy is essential for accurate diagnosis. The gland swellings of other reticuloses are very *similar, and biopsy, X-ray, and blood and marrow examinations may all be required for* their accurate definition.

Secondary carcinomatous glands of the neck are distinguished chiefly by their hardness. They occur in an age group which is not prone to, but is not immune from. tuberculous adenitis. Their presence, or a suspicion of their nature, should lead to search for a primary growth in the mouth, pharynx, chest, thyroid and stomach. These secondary growths may ulcerate through the skin and give rise to sinuses.

Branchial Cysts occur in the same situation as the earliest swellings in upper cervical adenitis and simulate the more chronic tuberculous abscesses. A smooth rounded swelling appears below and behind the angle of the jaw. Differentiation may be difficult. The chief points of distinction are that only rarely are outlying glands, quite incidental, to be found as are often present in tuberculous adenitis; the swelling, apart from when infection occurs in it, lacks the tension of a tuberculous abscess, being flaccid to the feel as if more fluid could easily be contained within its capsule; the aspirated fluid of a branchial cyst contains crystals of cholesterin easily demonstrable by a microscope.

Actinomycosis has its commonest site in the lateral cervico-facial region. Here, however, it is the skin and subcutaneous and deeper connective tissues that are affected, not the glands, and a brawny indurated area develops in which appear multiple sinuses discharging the sulphur granules of the ray fungus. A *carotid body tumour* is distinguished by its intimate relation to the carotid vessels and by the pulsation transmitted therefrom.

General examination of the patient must be carried out. In particular the chest should be X-rayed since a small proportion of the patients will be found to have pulmonary tuberculosis, a state which calls for distinctions in treatment. The Mantoux and Von Pirquet skin reactions are only likely to be helpful if negative: their negativity almost certainly rules out the diagnosis. It is rarely necessary, from a diagnostic point of view, to aspirate the abscess and examine the pus. Biopsy in the upper and lower cervical types, which both tend to caseation and softening, is to be avoided lest troublesome sinuses develop; in the diffuse form, as has been said, biopsy is almost an essential.

Treatment. Numerous forms of treatment have been advocated and it is difficult to assess their respective merits because of the undoubted tendency to spontaneous, if slow, subsidence of the disease. Broadly these treatments may be called medical, surgical, and radiotherapeutic. While they are commonly combined, the emphasis is usually on one or other of them. Medical treatment always implies improved general hygienic and dietetic measures; it may entail enforced general rest, local rest by immobilization of the head and neck, light therapy and the exhibition of drugs and antibiotics. Surgical treatment may take the form of evacuation of abscesses, extirpation of the gland masses, and

the removal of tonsils and adenoids. Radiotherapy, when used, is as an adjuvant to conservative medical treatment. Strong claims are made for its efficacy but they are difficult to substantiate in view of the likely spontaneous subsidence. The same reference may be made to various forms of light treatment.

Controversy has in the main been most marked between the advocates of conservative medical treatment, who reserve surgery for the evacuation of abscesses and the removal of tonsils, and the advocates of removal of the enlarged glands by dissection. The latter urge that the radical procedure should be performed as soon as the initial periadenitis has subsided, usually two or three months after the commencement of the disease.

The probability is that the correct care of any individual patient may follow either or both of these lines. Treatment for the first six months, at the minimum, should be conservative apart from the evacuation of abscesses. There is little sense, and perhaps much difficulty, in the dissection of glands before this elapse of time, or even much before an interval of nine months or a year. By then it will usually be clear whether the disease is subsiding spontaneously and by then also the natural resistance of the patient should have reached its highest. Removal of a mass of enlarged glands at too early a stage is quite likely to be followed by the disconcerting development of further similar swellings even directly under the scar. Quite a number of patients, however, still have an indolent mass of enlarged glands after nine months or more. These swellings vary a little in size from time to time but show no proper tendency to recession. It would seem that there is just enough resistance to keep them in check; indeed it would appear that they themselves are part cause of the lowered resistance and that their total removal can only do good. While this removal is not usually followed by recurrence, other masses may appear after the lapse of years.

The conservative treatment in the early stages will vary with the severity of the disease. When the mass appears in an acute form or where involvement of other groups of glands proceeds apace then both local and general rest are required. Sanatorium treatment may be needed for the worst as it will be also for the few whose general condition is poor and for those who show a chest lesion on radiography. The great majority, however, need not leave home and require little restriction of their activities. Local rest, to limit muscular movements of the neck and thus to slow the lymphatic stream and hinder spread of the infection, is obtained by the use of suitable splints or by bandaging. These should achieve limitation of both movements of the head upon the neck and the neck upon the thorax. There is, however, little advantage in such restriction when it is seen that the process is not spreading or that periadenitis has receded or in such patients in whom the process has been slow and chronic from the beginning.

The medical treatment during the conservative period consists in the usual measures for the building up of natural resistance, fresh air, plenty of good food and adequate rest. Vitamins are often prescribed. Calciferol is regarded by some as of value and given in doses of up to 50,000 units daily. It is highly doubtful whether the administration of streptomycin is likely to be of advantage except where there is coincident lung disease or in the uncommon instances where the disease is spreading rapidly and the masses are breaking down at many points in the neck.

Evacuation of Abscesses. It is essential that abscesses should be evacuated before they break through the skin of their own accord, when simple pyogenic infection will be admitted, a troublesome sinus may ensue, and the eventual scar is likely to be depressed

In *glandular fever* the swellings may be found elsewhere than in the neck, although sometimes they may be apparently confined to it. They are diffuse, discrete, tender and tense. Usually moderately enlarged, they may on occasion become as large as a plum; there is no tendency to suppuration. There is a mild intermittent fever, the blood shows a lymphocytosis and the diagnosis is established by the Paul Bunnell test. In *lymphadenoma* (Hodgkin's disease) the glandular swellings are of moderate size, soft to firm, smooth, discrete, and not tending to caseation; they simulate closely the diffuse form of tuberculous adenitis. Glands elsewhere are also commonly affected; the spleen is palpable in about one-third of cases. Intermittent fever (Pel-Ebstein) may occur. Biopsy is essential for accurate diagnosis. The gland swellings of other reticuloses are very similar, and biopsy, X-ray, and blood and marrow examinations may all be required for their accurate definition.

Secondary carcinomatous glands of the neck are distinguished chiefly by their hardness. They occur in an age group which is not prone to, but is not immune from, tuberculous adenitis. Their presence, or a suspicion of their nature, should lead to search for a primary growth in the mouth, pharynx, chest, thyroid and stomach. These secondary growths may ulcerate through the skin and give rise to sinuses.

Branchial Cysts occur in the same situation as the earliest swellings in upper cervical adenitis and simulate the more chronic tuberculous abscesses. A smooth rounded swelling appears below and behind the angle of the jaw. Differentiation may be difficult. The chief points of distinction are that only rarely are outlying glands, quite incidental, to be found as are often present in tuberculous adenitis; the swelling, apart from when infection occurs in it, lacks the tension of a tuberculous abscess, being flaccid to the feel as if more fluid could easily be contained within its capsule; the aspirated fluid of a branchial cyst contains crystals of cholesterin easily demonstrable by a microscope.

Actinomycosis has its commonest site in the lateral cervico-facial region. Here, however, it is the skin and subcutaneous and deeper connective tissues that are affected, not the glands, and a brawny indurated area develops in which appear multiple sinuses discharging the sulphur granules of the ray fungus. A *carotid body tumour* is distinguished by its intimate relation to the carotid vessels and by the pulsation transmitted therefrom.

General examination of the patient must be carried out. In particular the chest should be X-rayed since a small proportion of the patients will be found to have pulmonary tuberculosis, a state which calls for distinctions in treatment. The Mantoux and Von Pirquet skin reactions are only likely to be helpful if negative: their negativity almost certainly rules out the diagnosis. It is rarely necessary, from a diagnostic point of view, to aspirate the abscess and examine the pus. Biopsy in the upper and lower cervical types, which both tend to caseation and softening, is to be avoided lest troublesome sinuses develop; in the diffuse form, as has been said, biopsy is almost an essential.

Treatment. Numerous forms of treatment have been advocated and it is difficult to assess their respective merits because of the undoubted tendency to spontaneous, if slow, subsidence of the disease. Broadly these treatments may be called medical, surgical, and radiotherapeutic. While they are commonly combined, the emphasis is usually on one or other of them. Medical treatment always implies improved general hygienic and dietetic measures; it may entail enforced general rest, local rest by immobilization of the head and neck, light therapy and the exhibition of drugs and antibiotics. Surgical treatment may take the form of evacuation of abscesses, extirpation of the gland masses, and

last should be removed in 48 hours lest any resulting cross marks should mar the cosmetic result.

Dissection is not indicated in the lower cervical type where abscesses should be dealt with as described above; treatment in this type is essentially conservative as it is also in the diffuse non-caseating type.

and disfiguring. It is almost as important not to attempt premature evacuation of the abscess. A blush of the skin over the swelling signifies the optimum time for evacuation; most of the mass will by then have softened and more complete clearance will be achieved.

The various methods employed in dealing with these abscesses are: aspiration through a hollow needle, incision over the abscess with complete closure of the skin after evacuation of the pus, and evacuation through a small stab incision. The first of these methods is disappointing since the pus is often too thick to pass through the needle, the still semi-solid or solid elements of the mass are not removed, and frequent repetition of the procedure is often necessary, so that secondary infection is apt to be admitted and a sinus results. The second method, incision over the length of the swelling, is unnecessarily drastic as no more can be achieved by it than by the third method. The third method is the most satisfactory. A quarter-inch stab incision, in the line of a skin crease, is made into the abscess, and such contents as do not flow out are ejected by firm pressure of the thumbs on the adjacent tissues; much solid tissue is thus expressed; a single stitch is inserted to obviate secondary infection. It may sometimes be necessary to repeat the procedure once or twice as further softening takes place in a large mass. An alternative or auxiliary manœuvre to pressure by the thumbs is curettage of the interior, using a Volkmann's spoon. The spoon is passed through the stab incision and then guided through the hole in the deep fascia or sternomastoid muscle; gentle curettage, with the spoon facing successively in all directions, is then employed. Such curettage has been condemned because of the risk of opening into the internal jugular vein but this complication will not occur unless clumsy force is applied.

When *Surgical Removal* of the mass is performed, on the indications given above, there are certain points to be observed and borne in mind. The incision must follow a crease in the neck or lie parallel to one. More than one such incision is advised by some writers, to help removal of the larger masses. This should rarely be necessary; if a single incision be made long enough, flaps can almost invariably be turned up and down sufficiently to allow sufficient access. Hæmostasis should be achieved by the subcutaneous and sub-platysmal injection of saline-adrenaline solution of 1 in 150,000 strength (11 minims of 1 in 1000 adrenaline chloride solution to 100 ml. normal saline). After reflection of the flaps the deep cervical fascia is incised over the mass along the anterior border of the sternomastoid. The outer surface of the mass having thus been reached, the operator works round close to its capsule separating adjacent tissues from it, preferably by the opening out of the blades of a small curved Mayo's scissors. It is essential to remember that the internal jugular vein will be one of the structures to be separated; it is invariably collapsed over the surface of the mass and if the capsule be closely followed the vein will gradually show up and fill with blood. Usually the operator will move from one part of the mass to another as circumstances allow of easy dissection. The most difficult area is in the upper back part where a tongue of glandular tissue runs deeply. The spinal accessory nerve is in danger of injury here; it crosses the transverse process of the atlas to enter the anterior border of the sternomastoid muscle an inch below its origin, but if these points are remembered it is usually easy to identify it. Caseous and even purulent areas are occasionally entered into but need not deter the operator. The mass having been removed, hæmostasis is achieved, a small drain (which will be removed in 24 hours) is inserted, the deep fascia and platysma are approximated with interrupted catgut stitches and skin sutures or clips are used lightly to appose the skin edges. These

there is a group of small nodes which are very liable to be overlooked at operation, tucked in as they are deep to the anterior margin of the trapezius.

The nodes arranged along the spinal accessory nerve become continuous above with the internal jugular chain, on the deep aspect of the sterno-mastoid. At this level too they lie close to the outlying nodes situated on the mastoid process and below the occiput. They number anything from 5–10 and most of them cluster round the nerve just where it

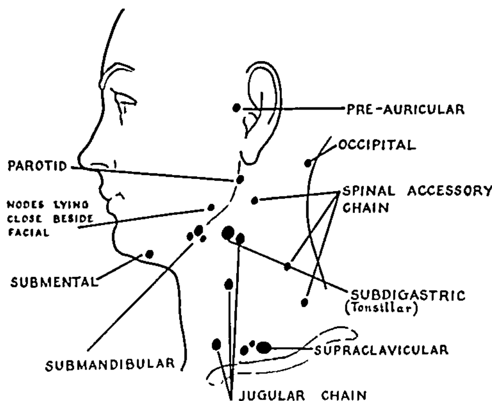


FIG 177 Diagrammatic representation of the principal groups of lymph nodes in the neck (after Rouviere)

emerges from the sternomastoid. Any attempt to conserve the nerve at this level during the course of a radical removal of neck nodes for cancer, is likely to make the operation an incomplete one and the same holds true for the branches of distribution of the cervical plexus at a slightly lower level. Included in their lymph catchment area is the naso-pharynx.

In addition to the three main groups, there are several other aggregations of nodes of varying importance. Chief among them is the group of 3–6 nodes, partly or completely under cover of the horizontal ramus of the mandible on each side. They are closely applied to the capsule of the submandibular salivary gland, in front of it, behind it and even an inconstant one in its substance. Two of the nodes are close to the facial vessels as they cross the lower margin of the jaw, where even in a healthy subject they can be rolled over the bone at the antero-inferior margin of the masseter. Resection of this group of nodes necessarily involves sacrifice of the salivary gland as well.

A second group of nodes is even more closely associated with the parotid gland which, because of its inaccessibility and of the closeness of its relations to the major vessels and

CHAPTER VI

NECK: CANCER IN THE CERVICAL LYMPH NODES. OTHER TUMOURS OF THE NECK. ACUTE INFECTIONS. CUT THROAT.

M. R. Ewing

CANCER IN THE CERVICAL LYMPH NODES

ANATOMY OF CERVICAL NODES

For a proper understanding of the surgery of cancer in the cervical lymph nodes (there is much to commend the adoption of "lymph node" in preference to "lymph gland." Martin has campaigned vigorously for the change and increasingly the former is finding acceptance in the American continent), we must have a complete knowledge of the disposition of the main lymphatics. Although subject to considerable variation, they follow for the most part a basic pattern.

The lymph from the entire head and neck is directed eventually into the great veins at the root of the neck, at the junction of the internal jugular and the subclavian. Two main drainage systems converge at this point (Rouvière, see Fig. 177).

The bigger and the more important comes from the row of nodes disposed along almost the whole length of the internal jugular vein. There are, however, seldom any nodes above the level where the vein is crossed by the posterior belly of digastric, so that it is widely held to be unnecessary to resect this muscle during a radical neck dissection. The nodes are intimately related to the vein and to its main tributaries, and are arranged partially on its lateral aspect, but mainly along its anterior margin. Of these the upper ones are the most frequently affected by metastatic cancer. Usually three in number, they occupy the interval between the posterior belly of the digastric, the internal jugular vein and its common facial or nearby lingual tributaries. The highest of these, and usually the biggest in the entire neck chain, is immensely important and is variously called the sub-digastric, jugulo-digastric or tonsillar lymph node.

A fairly constant node situated at a somewhat lower level in relation to the intermediate tendon of the omo-hyoid is often called the jugulo-omohyoid node and is reputed to be sometimes the first node to be implicated from a primary in the anterior part of the tongue.

Situated above the clavicle and arranged along the transverse cervical vessels (the artery usually lies deep to the nodes) is a second but smaller chain, which extends from the deep aspect of trapezius to the confluence of the internal jugular and subclavian veins. These nodes have important communications with the lymph drainage of the upper extremity and of the breast and the most medial one of the group is the node of Virchow. Into the outer end of this chain, moreover, there drains the lymph from a third and immensely important row of nodes which are intimately related to the spinal accessory nerve as it crosses the posterior triangle. At the point of confluence of these two chains,

to parotitis, or to any of the other benign causes which our wishful thinking may suggest to us.

(3) *The patient who comes to consult us on account of a neck swelling.* It is this last which constitutes the major diagnostic problem and to its resolution our subsequent remarks will largely apply.

The detection of a swelling in the neck poses a number of questions.

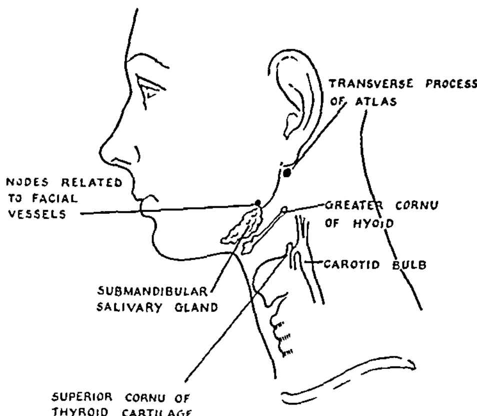


FIG 178. Diagram to indicate some of the normal structures in the neck which may be mistaken for enlarged lymph nodes.

(A) IS THE SWELLING A NORMAL STRUCTURE? (see Fig. 178)

(*a*) The tip of the greater cornu of the hyoid bone, situated as it is at the anterior border of the sternomastoid, 2·5 cm. below and slightly in front of the angle of the mandible may simulate quite closely an enlarged sub-digastric (tonsillar) lymph node. Firm, round and mobile, it is more likely to be confounding when in a stout patient it is well developed, than in a thin subject where its continuity with the remainder of the bone can usually readily be demonstrated. The easiest way of solving the problem is to feel for a matching swelling on the opposite side. Pressure over it, by dislocating the whole hyoid bone, will then usually suggest the real diagnosis.

(*b*) The superior cornu of the thyroid cartilage may similarly cause some difficulty.

(*c*) The transverse process of the arch of the atlas is palpable as a deep-seated firm swelling between the tip of the mastoid and the angle of the mandible. Firm pressure over it will demonstrate its unyielding fixation and its bony firmness and will cause acute discomfort.

nerves in the vicinity, does not readily lend itself to dissection. The most evident of these lie immediately anterior to the tragus and superficial to the aponeurotic investment of the gland. Others are disposed deep to the fascial covering on the lateral aspect of the gland or tucked between its lower pole and the sternomastoid, in the closest relation to the external jugular vein at its point of origin. There are other nodes which lie in the gland itself, usually closely applied to the vessels. At least some of the nodes in these last two sub-groups can be removed by amputation of the lower pole of the gland.

Between the diverging anterior bellies of the digastric, and bound down by a fascial investment to the mylohyoid, is a cluster of two, three or even more nodes of varying size which constitute the submental group.

The nodes which follow the anterior and external jugular veins are small and relatively unimportant and are in any event routinely sacrificed in the course of a radical neck dissection. The remaining cervical lymph nodes lie close to the structures which they drain, such as the pharynx, larynx, thyroid and trachea.

It should be remembered that although we are both clinically and at operation most impressed by the groups of lymph nodes, they are of course intimately connected one to another by numerous and freely anastomosing lymph channels. Any operation designed to resect the lymph drainage system of the neck must endeavour to achieve a block removal of the nodes and their connecting lymphatics. Any attempt to dissect the nodes alone is bound to be incomplete. Fortunately, the three main chains, ranged along the internal jugular vein, the trans-cervical vessels and the spinal accessory nerve are all enclosed in a continuous investing sheet of loose connective tissue. When the main jugular vein is separated from the carotid, the nodes come away freely with the vein. There is left behind little apart from a very naked looking carotid vessel and the accompanying vagus nerve.

THE CLINICAL PROBLEM

We may be confronted with the problem of cancer in the cervical nodes in a variety of ways.

(1) *The patient who is found to have, somewhere in the head or neck, a tumour which is known or believed to be a cancer.* Our routine examination involves a meticulous search of the entire regional node area. We must record and interpret our clinical findings in an attempt, either to clinch the diagnosis where it is still uncertain, or to assess the extent of spread of the cancer when this diagnosis has already been established.

(2) *The patient, who—having been treated for cancer—attends at a follow-up examination.* This again involves routine inspection of the regional nodes (or of the regional node area if they have already been treated by surgery).

The problem in these two groups is somewhat similar. We must be certain that, in *our routine examination, we review every likely site of a metastatic deposit* in a lymph node. We must remember the high risk of a contra-lateral metastasis when the primary is near to the mid-line, and the not inconsiderable risk of this occurring even in a strictly one-sided lesion, especially when it seems to have been adequately controlled on the side mainly involved. Every swelling, however small and however indefinite, which during any follow-up is noted for the first time, must be diagnosed and treated as cancer until it is proved to be otherwise. The same holds true for the lump which we believe to be inflammatory, to be due to irradiation, to unabsorbed suture material, to bone necrosis,

(D) If the Swelling is a Lymph Node, to what is its Enlargement Due?
(1) Is it Due to Infection?

Except when associated with a chronic skin infection, pyogenic lymphadenitis is usually easy to recognize. Infection is, however, often present along with metastatic cancer in neck nodes, especially when there is an ulcerated primary in the mouth. Under such circumstances, the cancerous neck nodes may present as an inflammatory swelling. The enlarged lymph nodes of a tuberculous infection are seldom so hard as those of metastatic cancer, and the later breakdown with abscess and sinus formation is characteristic. A mistake in diagnosis is most likely to be made when slowly progressive tuberculous cervical lymph node enlargement first presents in the adult.

(2) Is it Due to Cancer?

The commonest cause of cervical lymph node enlargement in the elderly is metastatic cancer. This provisional diagnosis should be made at the very outset and rejected only when it can confidently be proved otherwise.

Typically, a node invaded by cancer is firm, rubbery or densely hard, but when the tumour is rapidly growing it may be quite soft. It is also important to remember that—especially when a primary in the mouth is gross and ulcerated—necrosis and infection may lead to a complete breakdown of the regional lymph nodes, which then present as an acute inflammatory swelling. Failure to recognize this happening may lead the unwary to incise such a swelling which invariably allows a tumour to fungate out on the neck.

Although initially mobile, as growth advances they tend to become fixed, but this physical sign they share with lymph node enlargements due to other causes.

The rate of enlargement is widely variable, but when the history of a neck swelling goes back over a matter of years with little obvious advance in size, a diagnosis of cancer seems highly unlikely.

(3) Believing it to be Due to Cancer, is it Primary or Secondary? The second of these alternatives is in an adult the more probable, and should first claim our attention.

In answering this question we will be helped in a variety of ways:

The situation of an enlarged node will usually guide us to the site of the primary: a review of the patient's symptoms, helped maybe by the judicious use of leading questions, may equally direct us. Nasal obstruction, dysphagia, the sensation of a foreign body in the throat, or any change in the character of the voice would be especially significant in this respect.

If the swelling is sub-mental we would first look on the chin, or on the central part of the lip, or gingiva or in the floor of the mouth.

If sub-mandibular, we think of the tongue, the adjoining part of the floor of the mouth, the gingiva, the lips, cheek, palate, nose and antrum.

A primary in the naso-pharynx metastasizes, usually to both sides of the neck, to nodes high up in the spinal accessory chain or in the region of the mastoid process. The clinician is immediately impressed by the fact that they are situated both high and far back in the neck (see Fig. 179).

Similarly, node enlargement in the jugular chain will, according to its situation, focus our attention on the mucous lining of the mouth, pharynx, larynx or upper œsophagus, or on the thyroid.

(*d*) It is often exceedingly difficult to differentiate a firm node overlying the carotid bifurcation and transmitting the thrust of the vascular pulsation to the finger, from the prominent and thickened bulb of an arteriosclerotic vessel. There is no way of being quite certain and one is always happy when there is no pressing need for an immediate decision.

(*e*) It should be remembered that one can often in health feel normal lymph nodes. They may measure as much as 1 cm. across. This is certainly true of the sub-mental and sub-mandibular groups and of some of the upper components of the jugular chain.

(*f*) The sub-mandibular salivary gland is often mistaken for an enlarged lymph node. It is important to look out for a corresponding swelling on the opposite side. It is especially liable to become enlarged after the treatment of a cancer of the floor of the mouth or of the alveolus in the vicinity of the orifice of Wharton's duct. It is, however, equally important to bear in mind that because of the very close association between the salivary gland and the regional nodes, it is often impossible to establish to which of the two components a swelling in the sub-mandibular region may be due.

(B) Is the Swelling an Enlarged Lymph Node?

(i) Situation of Swelling. Although nodes are widely and variously scattered through the neck, it is obvious that one would hesitate to suggest the diagnosis of lymph node enlargement in a situation where none is commonly found: and, of course, the converse holds true.

(ii) Number of Swellings. If there is more than one swelling a diagnosis of lymph node enlargement is highly likely. Multiple swellings in the neck due to other causes are uncommon.

(iii) The Size of the Swelling is of no great help. It is obvious that when a lymph node is only slightly enlarged it will resemble closely in size a normal node: it is, however, equally important to bear in mind that a node involved by, say, a malignant lymphoma or by metastatic cancer (e.g. with a tonsillar primary) may reach massive proportions.

(C) If the Tumour is not an Enlarged Lymph Node, what Other Neck Swellings must be Considered in the Differential Diagnosis?

(1) Enlargement of the sub-mandibular salivary gland due to chronic sialitis or to a mixed tumour.

(2) A tumour in the region of the lower pole of the parotid gland, whether it be a mixed tumour or an adenolymphoma.

(3) A branchial cyst.

(4) Carotid body tumour. This is a clinical rarity, which presents as a firm, slowly-growing and painless tumour, just below the angle of the mandible. The diagnosis is commonly established only at exploration or on subsequent section.

(5) Neurilemmoma. This is another rare, relatively painless and slowly-growing tumour in the neck, which runs a benign course. Like the carotid body tumour it is seldom recognized before operation.

(6) A thyroid adenoma, especially if it is situated far laterally in the gland.

the patient to push the tongue out as far as it will go. If the tip is then gripped firmly with a piece of gauze and pulled over to the side away from the lesion and the corner of the mouth is simultaneously retracted on the same side, this part of the tongue can in most patients be freely exposed.

(c) *Cancer of the base of the tongue.* This part of the tongue is quite inaccessible to ordinary physical examination. It is in this situation too that there occurs not infrequently a rounded lesion which is non-ulcerated and easily overlooked.



FIG 180 Neck swellings of 2 years' duration due to lymph node enlargement (Diagnosis—chronic lymphatic leukæmia)

(d) *A cancer of the tonsil,* while small, may give few symptoms and here again it is only too readily overlooked.

(e) *Cancer of the vallecula,* may also be the site of a "silent" primary.

(f) *Cancer in the pyriform fossa or adjoining hypopharynx* may be, in its early stages, completely latent and present first as a neck swelling only when it has metastasized to the regional nodes. A tumour here may only be seen on direct examination, as will a small cancer at the upper end of the œsophagus.

Having examined with the greatest of care every nook and cranny in the mouth, pharynx and larynx, we must now direct our attention elsewhere.

Deposits in the supra-clavicular nodes make one think of a bronchial primary, or even of a cancer below the level of the diaphragm. Careful examination of the arm and of the breast and axilla would also be obligatory.

Detection of the primary is something that may be perfectly obvious: alternatively, it may exercise our diagnostic acumen to the limit. It is so easy to miss a small cancer, even in the accessible areas of the mouth.



FIG 179. Metastases in the upper nodes of the spinal accessory chain from a primary in the nasopharynx.

Among those which experience has shown often to be overlooked are the following:

(*a*) The most elusive primary of all is a *cancer in the nasopharynx.* Typically small, and sometimes even of microscopic proportions, it is commonly silent. Inaccessible to direct inspection, and elusive even to the experienced endoscopist, it is often missed for a long time until the later development of cranial nerve palsies makes the diagnosis all too evident.

(*b*) *Cancer of the side of the tongue close to the base of the anterior pillar.* Some patients find it exceedingly difficult to allow any adequate examination of this area, and resist strenuously with the tongue and involuntary gagging Access is facilitated by asking

increasing frequency, by what are often bizarre swellings, which further investigation proves to be the distant metastases of an unsuspected lung primary. Enlargement of the lower cervical lymph nodes is often the first symptom. This diagnosis especially suggests itself in a middle-aged male who smokes: the presence of finger-clubbing is a valuable piece of corroborative evidence.

(*j*) *Breast.* An enlarged supraclavicular node is occasionally the first sign of a breast cancer, especially when the primary is either wilfully overlooked or masked by a thick layer of investing fat and breast tissue.

(*k*) *Stomach and other abdominal viscera.* Although enlargement of Virchow's node (Troisier's sign) is occasionally a sign of a visceral cancer below the level of the diaphragm, it is unusual for it to be the first one. An abdominal neoplasm must, however, be remembered as the possible source of a supraclavicular node metastases.

A careful clinical review having been made of every possible local and distant source without the discovery of any primary cancer, we are now driven to the diagnosis of a primary malignancy in the lymph nodes, such as Hodgkin's disease, lymphosarcoma or any of the other reticuloses (Fig. 180). To establish such a diagnosis we must look carefully for lymph node enlargement elsewhere and for enlargement of the liver and spleen, for alterations in the blood picture and X-ray signs in the lung and mediastinum.

It is at this stage in our investigations that we are usually forced to do a biopsy to establish the diagnosis.

Most often this is done as a formal surgical excision of a readily accessible node.

The alternative of some method of needle or of aspiration biopsy has its firm adherents. Provided the technique is mastered and the pathologist is accustomed to the handling of small biopsy specimens, and always provided too there is an awareness of the limitations of this method, it offers a very easy way in a high percentage of cases of arriving at the diagnosis quickly and with the minimum upset. It must be admitted, however, that the diagnosis of the cause of lymph node enlargement is one of the most difficult with which the pathologist is confronted.

The diagnosis may, by this manœuvre, be firmly established by the pathologist, but when the biopsy reveals metastatic squamous carcinoma, the problem is thrown back once again to the clinician. In former times the presence of a squamous epithelial tumour in the soft tissues of the neck in the absence of a recognizable primary, was the justification for the diagnosis of "primary branchiogenic cancer," on the assumption that such tumours originated in remnants of the branchial clefts. Such a diagnosis is now no longer acceptable: the experience of diligent clinical examination and re-examination and of assiduous follow-up and the findings of painstaking dissection at autopsy have shown that in the vast majority of cases a primary can be demonstrated. It may measure only a few millimetres across and will elude all but the most careful enquiry. It can be taken for granted that the number of cases in any clinic in which the diagnosis of branchiogenic cancer is made, varies inversely with the care with which the search for a primary is conducted.

What then are we to do when the biopsy report on a resected node reveals metastatic cancer and when the primary is unknown? The diagnosis must remain open and every possible site examined repeatedly and with consummate care until the site of the primary is established. Unhappily, it may never be established during life and it may remain undetected even at autopsy.

(*g*) *Thyroid.* Papillary thyroid cancer often first presents as a laterally situated swelling in the neck—the so-called "lateral aberrant thyroid" of former times, now known to be a lymph node metastasis from a small and often overlooked primary in one lobe. The whole length of the jugular chain of lymph nodes may be involved, and the history is often a long one. Tuberculous lymphadenitis is the diagnosis which in this country



FIG 181. When this patient reported with enlarged lymph nodes below the right mandible, the small pigmented tumour on the lid margin passed unnoticed. When it was later excised it was found to be a *malignant melanoma with lymph node metastases.*

first suggests itself, but it should be borne in mind as an infrequent cause of widespread lymph node enlargement in the young.

(*h*) *Malignant Melanoma.* Malignant melanoma spreads typically by lymphatics to the regional nodes. It is the easiest thing in the world to overlook in the investigation of a patient with a neck swelling, a small, insignificant and symptomless pigmented skin [illegible] (see Fig. 181). It is important to explore the skin of the arm [illegible]

of the scalp with the aid of a [illegible]

(*i*) *Bronchus.* Physicians and surgeons alike are being confronted, with an [illegible]

the platysma. The sterno-mastoid, omohyoid and at least the anterior belly of digastric must be sacrificed as well as the whole length of the jugular vein and its tributaries. Attempts to retain the eleventh cranial nerve compromise the completeness of the resection, but care should be taken to preserve the vagus, the phrenic, and the hypoglossal and lingual nerves, and the common and internal carotid vessels, with the sympathetic trunk.



FIG. 183 Post-operative photograph of a patient who had, in two stages, a total glossectomy (with sacrifice of the entire mandible) and a bilateral "block dissection" of neck.

The operation can quite safely be done on each side (Fig. 183). It would seem reasonable when there is bilateral involvement to operate first on the side most heavily invaded. It is the practice in this country to sacrifice the second internal jugular only if an interval of 3 months or so has passed since the first operation. It seems, however, that there is no need to be fearful of the results of resection of both jugular veins even when a simultaneous bilateral operation is carried out. Suprahyoid and other modified or partial neck dissections are unsatisfactory: it is preferable always to carry out the complete operation.

TREATMENT OF METASTATIC CERVICAL CANCER

Surgery

It is generally admitted, even by radiotherapists, that cancer in the cervical lymph nodes is best dealt with surgically, by a block or radical neck dissection. "Dissection" is



FIG. 182. A double-Y incision gives excellent access in doing a neck dissection and lessens the risk of necrosis of the flaps

in some ways an unfortunate term, since it implies a removal of cancerous lymph nodes in the same way as for tuberculosis.

Scope of Operation. An adequate operation is planned to remove, as a single block of tissue, all the lymph-bearing tissue from the mandible above to the clavicle below and from the midline in front to the trapezius laterally. A variety of incisions has been described: the double-Y, as used in the Memorial Hospital in New York (Martin *et al.*, 1951) and based on a wide experience affords excellent access with the minimum risk of flap necrosis (see Fig. 182). The practice varies in relation to the retention or sacrifice of

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certainly no bar to a satisfactory excision. The operation is not a shocking procedure and even the old and feeble can be submitted to it without misgivings.

When the regional nodes are enlarged and mobile their removal is obviously obligatory: it is never allowable to wait to see if their enlargement is due to infection—it so seldom is. Surgery is undertaken only when treatment of the primary is complete: some prefer to wait until it is healed

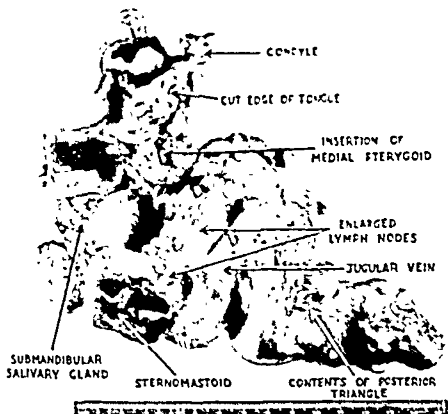


FIG. 185. The tissue removed in an "en bloc" excision of a cancer of the tongue with cervical lymph node metastases.

In the absence of detectable regional lymph node enlargement practice varies in different clinics. Those who practise in such cases immediate operation (the so-called "prophylactic" procedure) would argue that just because the nodes cannot be felt there can be no guarantee that they do not contain cancer, and that surely the best time to do a neck dissection is at the earliest possible moment. Further, they would say that high frequency of spread to lymph nodes from a primary, say, in the tongue, makes immediate operation the only reasonable practice.

Those who adopt the expectant treatment and who embark on block dissection only when there is clinical evidence of lymph node enlargement, believe that by so doing they save a good many patients an unnecessary operation, and that by waiting they do not appreciably lessen the patient's prospect of cure. In the case of a lip primary where, in an early case, there is no great risk of spread to the lymph nodes, the waiting policy is clearly justified, but with lingual cancer where there is a very high percentage of cervical

Disability Following Operation

The disability following radical neck dissection is inconsiderable. Loss of the accessory nerve is adequately compensated for. Weakness of the depressor of the lower lip (see Fig. 184) and œdema of the lower part of the face are seen very frequently (Ewing and Martin, 1952).



Fig. 184 Patient with mouth open to demonstrate paralysis of the depressor of the right lower lip from damage to the mandibular branch of the facial nerve during the resection of a large dental cyst

Indications

Before we embark on surgery, we can assume that it is worth considering only when the primary has been, or is likely to be controlled, either surgically or by irradiation. It is equally apparent that there should be no distant metastases. Fixation of the involved nodes, which is so often given as a contra-indication to operation, is a difficult point to determine clinically, and cannot be accepted as a complete bar to resection. Every structure in the neck can reasonably be sacrificed in the course of a block dissection excepting only the carotid vessels and the vagus. Involvement of skin or of bone is

The proper management of the case where the diagnosis remains "metastatic cervical cancer: primary undetermined" is always somewhat unsatisfactory. Most often the neck is irradiated and it is true this is occasionally followed by cure, the presumption being that the undetected primary has been included in the treated area. A case can also be made for doing a block dissection in such cases, especially if the node enlargement is a bulky one, reserving radiation for control of the primary when it is eventually discovered. It can then be used with precision and in high dosage without any risk of radionecrosis.

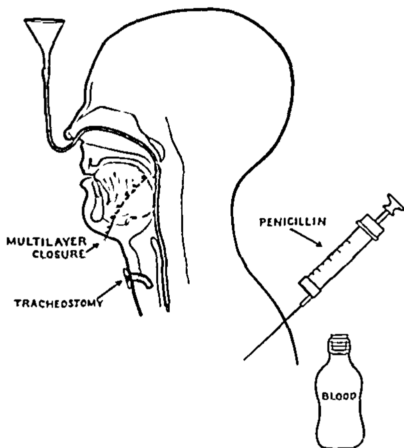


FIG 187

Resection of the primary with simultaneous block dissection. It seems somewhat unfortunate that it has so long been the practice in the surgery of mouth cancer to consider the management of the primary as a distinct and separate problem from its control in the regional lymph nodes. This practice was no doubt determined to begin with by the problems and hazards involved in a resection involving not only the oral cavity and the cellular planes of the neck, but the intervening bony framework of the jaw as well. Hesitation in attempting this type of operation was also encouraged by the clinical observation that in the average case recurrences did not seem often to be encountered between a primary, say, in the tongue and in the regional lymph nodes.

There has, however, been manifest especially in the United States, a great interest in *en bloc* operations planned to include the primary in the mouth, the lymph nodes in the

metastases, there is room for difference of opinion. Those who counsel the prophylactic operation should, in the case of a tongue primary, practise routine bilateral neck dissection, especially if the tumour is centrally situated, or is at the base or has transgressed the mid line.

Adoption of an expectant attitude—and, by and large, this is the usual practice in



FIG 186 One way of extending the standard incision for radical neck dissection so that a cheek flap can be turned back to give access to the jaw and to the mouth

most clinics in this country—throws a great load of responsibility on the clinician. Such a line of treatment is acceptable only if the surgeon and the patient alike can be trusted to maintain an unbroken follow-up at regular intervals. In a community where continuity of supervision cannot be assured the expectant programme has no place. At each attendance there must be a meticulous examination of the neck (remembering always the risk of contralateral metastases), preferably by two examiners. The detection of any asymmetry, or of any swelling, however small, which was not noted at an earlier examination, must be taken as evidence of metastatic spread and made the reason for prompt operation.

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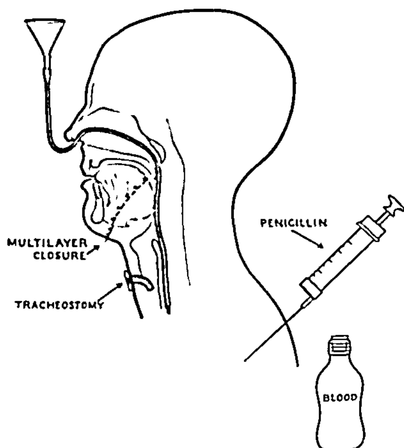


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neck, and all the tissue between the two (Fig. 185). To resect an alveolar tumour, or a cancer of the margin of the tongue, it is necessary to excise a segment of the mandible, partly to gain proper access, but mainly to make possible a primary reconstruction of the mouth. This can conveniently be done by prolonging the anterior part of the neck incision to split the lip in the midline (Fig. 186). It is, for this purpose, necessary to open up the tissue planes in the sub-mandibular triangle. It would seem in such an event to be

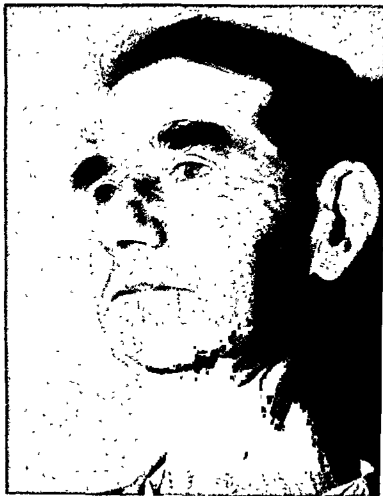


FIG 188 Post-operative photograph of a patient of 68 who 5 months previously had a resection of a major portion of the tongue, the left half of the mandible and a simultaneous "block dissection" of the neck

a wise and reasonable extension of the resection to go ahead to do a simultaneous block dissection, whether there is clinical evidence of regional lymph node enlargement or not. Experience has shown that operations of this type can often be carried out even in the elderly with a low mortality and morbidity. Every effort must be made to achieve a one-stage reconstruction of the mouth without the need of recourse later to a complicated plastic repair. It is essential to get primary healing by meticulous multi-layer suturing. A tracheostomy is a wise precaution for the first few days and the patient's nutrition can be maintained by a high calorie diet given by an œsophageal tube (Fig. 187).

The cosmetic and functional result following operations of this type is usually quite acceptable (Fig 188), although difficulties in masticating a full diet are often insurmountable.

En bloc operations of this sort are applicable also to the management of laryngeal, parotid and thyroid cancer.

Irradiation

Although surgery is the method of choice in the treatment of metastatic cervical cancer, it obviously has its limitations.

It is not possible to give a cancericidal dose of radiation to the whole lymphatic field, even on one side of the neck. Radiation can, however, be sanctioned under the following circumstances:

(1) When the primary is inoperable. This would obtain, for example, when the primary is in the posterior third of the tongue or in the tonsillar region. It would seem reasonable under such circumstances so to place the radiation fields in relation to the primary as to include the neighbouring lymph nodes. When there are bilateral metastases there would be an even stronger call for radiation, especially as the nodes in such cases are often bulky, highly anaplastic and often strikingly radio-sensitive (although not radio-curative).

(2) When the primary is uncontrolled, or where experience suggests that it is uncontrollable, palliative irradiation has some merit.

(3) It can be used as a palliative when the nodes are either hopelessly fixed clinically or found at operation to be seriously compromising the main artery, as well as in the prevention and alleviation of the distress which comes with fungation of the cancer out on to the skin of the neck.

(4) When the patient is unwilling to consent to surgery, or where, alternatively, his general condition is unfit for it. Age alone is, however, no bar to block dissection.

(5) There is seldom any alternative to irradiation in the treatment of a local recurrence following a block dissection.

(6) When no adequate surgical facilities are available.

Treatment which involves surgery, combined with pre-operative or post-operative irradiation (or both), has not found much acceptance in this country.

Prophylactic irradiation in the absence of clinically enlarged nodes has no merit.

OTHER TUMOURS OF THE NECK

The clinical problem of a neck swelling is always an intriguing one. Most often cancer features largely in one's consideration and most of the common swellings have been dealt with under the differential diagnosis of this disease. A few of them will, however, be considered here at greater length

CYSTIC HYGROMA COLLI: LYMPHANGIOMA

The neck is the most common site of cavernous lymphangiomata. The great majority of these are seen in infancy or early childhood. A large tumour may be noticeable at birth. It may disappear spontaneously. More often, however, it increases in size slowly or quite rapidly. This latter happening is often the result of infection occurring

spontaneously possibly from the tonsil or following an attempted surgical removal. Alternatively it may gradually increase in size until it reaches very large proportions.

A swelling is noticed in the neck which is often very conspicuous and which is soft, bulky, diffuse and compressible. Its main bulk is situated in the subcutaneous tissue and transilluminates. The tumour is usually situated in the posterior triangle somewhere above the clavicle where it can cause symptoms by pressure on the cervical or on the brachial plexus but it may occur at a higher level in the anterior triangle or in the vicinity of the floor of the mouth. Urgent respiratory difficulties may result in such cases.

The tumour is not a neoplasm in the true sense but is due to a developmental anomaly. It is usually multiloculated and consists of a conglomerate of thin-walled cystic spaces, each containing clear fluid. It will often be found to have spread widely among the adjoining muscles and may also have ramifications which extend as far as the base of the skull above and the axilla and mediastinum below. Histologically these are dilated lymph spaces with lymphoid tissue in the wall. Often too it contains many large cavernous blood spaces.

Operation

Every effort should be made to postpone operation as long as possible. The technical difficulties of the removal of the tumour in early life when the normal anatomical structures are small are very considerable and attempts at excision early in life are associated with a prohibitive mortality, and often too by the disappointment of recurrences. Treatment will be instituted only in the presence of rapid growth and pressure symptoms. A simple incision should be avoided.

If operation ultimately becomes necessary its complete eradication may involve a long and tedious operation. This should only be undertaken under ideal conditions and through a generous incision.

Clinical experience seems to suggest that in addition to the multi-locular and diffuse lymphangiomata there is another variety consisting of only a few large cystic spaces. This is the kind that reacts well to the injection of a sclerosant such as sodium morrhuate and equally its surgical excision is a relatively easy problem.

Removal of the lymphangioma illustrated in Fig. 189 presented no difficulties. It was possible to remove it intact, in its entirety, by a wide exposure and patient dissection.

CAROTID BODY TUMOUR

The carotid body is a very small ovoid structure firmly attached to the posterior aspect of the carotid bifurcation. It is generously supplied with vessels and is sensitive to certain changes in the chemical composition of the blood. A tumour which arises in this structure is such an uncommon neck swelling as to be considered to be a rarity. Known to earlier generations of surgeons as the "potato tumour of Hutchinson" it is now increasingly the fashion to call it a "chemodectoma" and to bracket it with the tumours of the other chemoreceptor organs such as the jugular body (glomus jugulare), vagal body and the aortic body.

The patient is commonly in his thirties, forties or fifties and usually comes seeking advice only on account of a neck swelling which is causing some disfigurement and which is very rarely bilateral. A striking feature of the tumour is the slowness of its

growth; in the average case it has been present for 5 years or more before the patient seeks advice.

Pain is never a prominent feature and pressure symptoms are similarly infrequent. On examination the tumour is found to be a hard ovoid or slight lobulated tumour



FIG 189. Cystic hygroma of neck. It contained only a few loculi and was removed intact without difficulty

deeply situated in the neck, partly under the sternomastoid muscle at about the level of the hyoid behind and often burrowing deep to the angle of the mandible. It will not move on swallowing. The tumour is so closely related to the bifurcation of the common carotid artery that it is often exceedingly difficult to decide whether the tumour itself is pulsating or whether it is only transmitting the pulse of the underlying vessel. Its deep attachment may be demonstrated by bi-manual examination, with one finger on the neck and the other in the pharynx. Dysphagia is an unusual symptom but when it is present there will usually be a medial bulging of the pharyngeal wall. It is always said that one of the typical features of a carotid body tumour is that it can be moved in a side-to-side direction but not vertically, but of the validity of this test there is conflicting evidence.

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Operation will be necessary only

(*a*) If the biopsy shows the tumour to be malignant

(*b*) If the tumour is causing pain or is growing towards the pharynx and causing dysphagia

In all other such cases, and they will form the great majority, it is enough to establish the diagnosis and to postpone consideration of a dangerous resection until there are urgent symptoms



FIG. 190 Lipoma of the neck

NEURILEMMOMA

This is an uncommon tumour which may take origin from any of the nerves in the neck. Most often they are deeply situated close to the side wall of the pharynx. The only one we have encountered was in this situation and appeared to be taking origin in the vagus nerve. The patient presents with a painless neck swelling and it is seldom possible to make a firm clinical diagnosis. It simulates closely a carotid body tumour and its nature is apparent only when it is explored at operation or when it is later examined by the pathologist.

There is a distinct risk of local recurrence and resection of the tumour along with the

Moving of the head may more rarely give rise to dizziness and even syncope and handling the tumour may precipitate similar symptoms (carotid sinus syndrome). Clinical evidence of recurrent laryngeal or of cervical sympathetic paralysis is most unusual.

Differential Diagnosis

This includes a long list of laterally situated neck swellings but in practice the issue usually narrows down to the consideration of a branchial cyst or a neurilemmoma or Schwann-cell tumour. A branchial cyst is not nearly so deeply placed in the neck and tends to bulge outwards. It seldom grows deep to the angle of the jaw and is much more mobile. The neurilemmoma is also a deeply situated tumour and one which grows from any of the nerves near the carotid bifurcation and will be quite indistinguishable clinically from a carotid body tumour.

Treatment

The clinical diagnosis of uncommon neck swellings is usually quite fallacious. It seems reasonable to advise operation in the first instance to establish a diagnosis. The incision should be a generous one. The tumour will be met with as an exceedingly vascular greyish-red tumour intimately bound down to and even grooved by the bifurcation of the carotid artery. It will bleed vigorously even on gentle handling, but may noticeably diminish in size on firm pressure proximally over the common carotid artery. The taking of a biopsy is not without some risk but it is a justifiable one. Having established the diagnosis of carotid body tumour treatment becomes a matter of opinion.

If the tumour can, by careful dissection, be peeled off a major vessel, this is the procedure of choice. Technically, however, it may be exceedingly difficult and division of the intrinsic vessels of the tumour which communicate directly with the lumen of the underlying major arteries may give rise to urgent and embarrassing hæmorrhage. Under such circumstances the main vessels should not be sacrificed for the bleeding can usually be controlled by suturing. It has been suggested that even the most firmly anchored tumour can be resected without ligating the main vessels by carrying out the dissection through the walls of the vessel and removing only the adventitia.

It is probably seldom justifiable to attempt to remove a carotid body tumour if this would involve the resection or ligation of the internal carotid artery or of the carotid bifurcation For this view there are several good reasons:

(1) Carotid body tumours grow very slowly, seldom give rise to urgent pressure or other symptoms and, if not molested, will not often be a cause of death.

(2) Malignant carotid body tumours are exceedingly rare.

(3) Attempts at removal which involve the sacrifice of the bifurcation have a prohibitive mortality as high as 30 per cent and, even among the survivors, there is a very high incidence of disabling paralyses This is the case even when every known manœuvre is carried out to test the response of the cerebrum to carotid occlusion and when every device is employed to improve the chance of its survival. The techniques which have been developed within more recent times, by which efforts are made to maintain the cerebral circulation by some form of vessel grafting are exacting and uncertain, even when supplemented by hypothermia and by the use of some temporary arterial by-pass (Lahey, F. H., Warren, K. W. (1951), *Surg. Gynec. Obstet.* **92**, 481)

cases there may be gas in the tissues due either to the visceral injury or to a gas-producing organism. Treatment is by free drainage and the proper treatment of the primary cause.

(7) Pyogenic osteomyelitis of the spine may give rise to an abscess which tracks laterally in precisely the same way as does a tuberculous spondylitis behind the prevertebral fascia and along the cervical nerve roots to present in the posterior triangle behind the sterno-mastoid. The abscess should be drained along the same tissue planes.

(8) The acute retropharyngeal abscess of childhood presents as a bulging of the posterior wall of the pharynx and is incised through the mouth.

(9) It is very important to bear in mind that lymph node metastases in the neck, especially from a necrotic and fungating primary, often becomes secondarily infected and present as an abscess. The unwary may overlook the diagnosis: incision is followed invariably by fungation of the tumour on the neck.

CUT THROAT

In civilized communities this is almost always a self-inflicted wound. Its seriousness depends on the depth of the cut rather than on its length and also the promptness with which treatment is instituted.

The greatest threat to life comes from hæmorrhage. Division of the carotid vessels or of the internal jugular vein causes very urgent bleeding which, if unchecked, will rapidly prove fatal. Fortunately these major vessels often escape since they are deeply situated in the neck and are protected in some degree by the sternomastoid muscles. The thyroid vessels are, however, more vulnerable.

The cut in the case of a right-handed person begins on the left side, where it is deepest, and inclines downwards and to the right. The external jugular vein on left side is often divided. Bleeding from this vein and from the skin vessels is always copious but, although the patient's clothing bears the evidence of considerable blood loss it has often stopped spontaneously by the time the patient comes for treatment. There is by the nature of things, often a long delay between the time of wounding and treatment. The wound may penetrate the larynx or pharynx—here injury is often below the level of the cords. A deep cut may result in considerable bleeding into the air passages with the immediate risk of respiratory obstruction and death from asphyxiation and the later hazard of lung collapse and infection. Injury to the air passages may be apparent clinically from the laboured and noisy breathing or from the presence of frothy blood at the site of injury.

Treatment

The patients are usually quite unco-operative and are occasionally completely negative in their attitude. Urgent bleeding will be arrested by digital pressure applied over a gauze on the bleeding point. Cyanosis will be a call for immediate tracheostomy or intubation and for vigorous bronchial aspiration.

In the less urgent case time can be spent before operation in blood replacement. If there is clinical evidence of injury to the larynx or trachea a formal tracheostomy should be fashioned and the anæsthetic administered through a cuffed tube. It is essential to have a good airway: bleeding is otherwise likely to be embarrassing. The wound is then explored, bleeding points being secured as they are encountered. An incised wound of

related nerve seems to be justifiable even although in the case of the vagus nerve this would lead to a cord paralysis.

LIPOMA

Although the most frequent site of a lipoma is on the back of the neck where the diagnosis does not usually present much difficulty, they are occasionally met with in the loose tissues on the side of the neck and especially just above the clavicle (Fig. 190). They differ in no way from lipomata at other sites, and have the same distinctive softness, lobulation and mobility.

ACUTE INFECTIONS

The skin of the back of the neck, especially in the vicinity of the collar line in men, is one of the commonest sites both of boils and of carbuncles.

(1) **Ludwig's angina.** Although this term is often used to describe any acute infection in the region of the floor of the mouth, it should properly be restricted to those cases where the infection involves the tissues above as well as those below the muscular diaphragm which forms the floor of the mouth. In this form it is fortunately now distinctly uncommon. The infection comes usually from a tooth and is characterized by the rapidity of its progression. There is alarming swelling and œdema of the floor of the mouth, so that the tongue is displaced upwards and eating and breathing become difficult. At the same time there is œdema below the jaw with redness of the overlying skin. There will be marked toxæmia and a high fever. The seriousness of this type of infection is now much less than it was in the days before the advent of antibiotics. Prompt treatment will usually bring speedy relief, but incision of the swelling may still occasionally be necessary. Pus formation is unusual; all that is required is an exploration (by Hilton's method) and drainage of the tense œdematous tissues.

(2) **An indolent abscess** below the jaw, which eventually discharges on the surface and leads to the development of a chronic discharging sinus, firmly fixed to the subjacent bone is usually found to be due to the insidious extension of a dental apical abscess through bone and soft tissues. It will heal promptly as soon as the offending tooth is removed.

(3) **Submandibular sialitis** due to calculous obstruction of the duct or more commonly to the presence of a stone in the gland itself usually lead to the development of an abscess round the gland. It may then be necessary to remove the gland in two stages, the first operation being limited to drainage of the abscess.

(4) Abscesses of the neck are not uncommon in childhood. The infection comes commonly from impetigo of the skin of the face or scalp, often associated with pediculosis capitis. It is important to remember that the need for the evacuation of pus is no less necessary now than it was in the days before the use of antibiotics. Healing occurs promptly so soon as the thick purulent content of the abscess is evacuated.

(5) The extension of infection from air cells through the tip of the mastoid process into the digastric triangle (Bezold's mastoiditis) was never common and must now be a rarity.

(6) A dangerous spreading cellulitis of the neck may follow operations on the œsophagus, or perhaps more commonly in association with an impacted foreign body or as the result of injury following an instrumentation (e.g. gastroscopy). Occasionally in such

CHAPTER VII

DISEASES OF THE BREAST

R. S. Handley

EMBRYOLOGY AND SURGICAL ANATOMY

Embryology. The breast develops from the milk line, a fœtal thickening of the ectoderm which runs down the ventral surface of the body from the axilla to the middle of the inguinal ligament. In the human fœtus, this line normally atrophies except in the pectoral region where a localized area proliferates to send buds into the mesoderm. These buds, some twenty or more in number, are destined to form the lobes of the breast by proliferation, by the formation of branching processes and, at about the eighth month of fœtal life, by canalization. At birth, the nipple is a shallow pit, into which the canals formed by the primary buds open, and not until puberty does it grow into a recognizable elevated nipple. Not only the ectoderm, but also the mesoderm take part in forming the breast, the latter arranging itself as ensheathing and supporting tissue for the ectodermal glandular elements.

Anatomy. The external appearance and limits of the breast are too familiar to require comment.

Secreting Tissue. The breast is a more extensive organ than its gross appearance suggests. The axillary prolongation or axillary tail of Spence is a well known and often palpable prolongation, but outlying tubules may also occur as high as the clavicle, as far medial as the midline, at the costal margin, and as far lateral as the mid axillary line. A simple mastectomy is thus a more extensive operation than the procedure generally passing under that title. The secreting tissue of the breast is arranged in lobes, twenty or more in number, each having a main duct which dilates into an ampulla just before it opens, separately from its fellows, on the apex of the nipple. Large branches of the main duct drain lobules, and divide into smaller and smaller branches as they are traced peripherally, until they end in the acini. The lobes of the breast cannot be separated by dissection because elements of adjacent lobes interdigitate and there is no regular surrounding capsule to afford a plane of cleavage.

Supporting Tissue. The fibrous tissue which supports the breast is of two varieties. There is tough white fibrous tissue which surrounds the breast and sends strong but irregular septa through its substance. The forward prolongations of these septa to the dermis are known as Cooper's ligaments (ligamenta suspensoria). There is also the delicate supporting periductal and periacinar connective tissue. Cooper's ligaments and the fibrous tissue septa join the pectoral fascia to the skin and are important in producing many of the clinical signs in breast disease by their deep and superficial connection. The breast also contains a varying but often large quantity of fat, much of which lies anterior to the secreting tissue.

The Nipple. The nipple, a firm dark pink structure, should normally protrude. It contains smooth muscle by means of which it can undergo a sort of erection when it is

the internal or of the common carotid artery should be repaired if at all possible using a continuous layer of arterial silk.

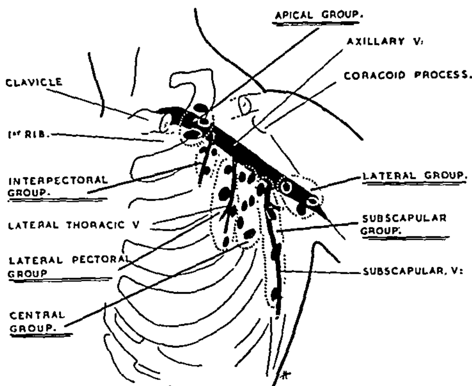
Injuries to the air or food passages are carefully sutured by interrupted sutures of catgut, the wound being closed with drainage. A gastrostomy is a wise precautionary measure when the pharynx or œsophagus has been incised.

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stimulated by suckling. It is surrounded by the areola under which are very numerous sebaceous glands; these are sometimes the seat of inflammation.

Blood Vessels. The breast receives most of its blood through the perforating branches of the internal mammary artery. A large supply also reaches it from the lateral thoracic artery. The subscapular, segmental intercostal, pectoral branch of the acromio-thoracic and superior thoracic vessels supply smaller branches. There is usually free anastomosis between these vessels. The veins in general follow the arterial pattern.

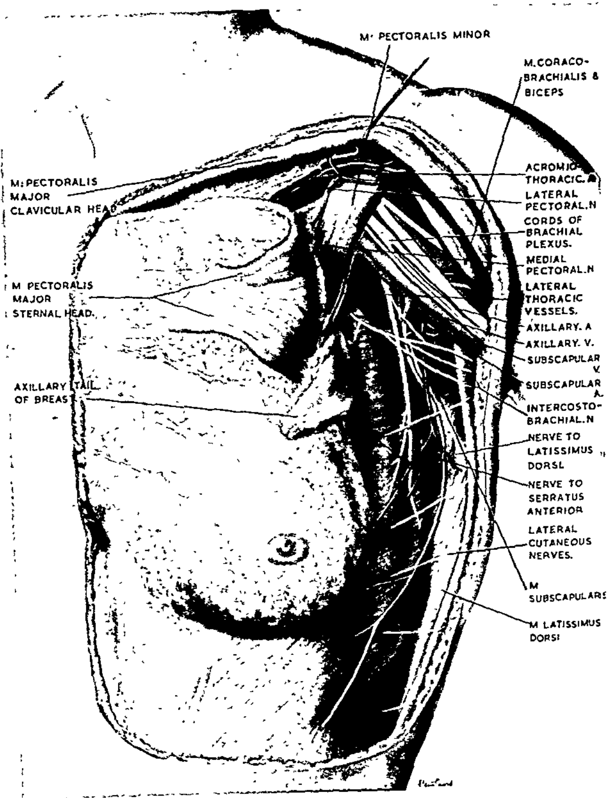


(From the author's article in Lewison's "Breast Cancer," by kind permission of Dr. E. Lewison and the Williams and Wilkins Company)

FIG. 192. Diagram of the axillary groups of lymph nodes which drain the breast.

Nerves. The nerves of the breast are derived from the first to the fifth thoracic nerves which beside transmitting afferent sensory stimuli, also send sympathetic motor fibres to blood vessels, sweat glands, and hair. There is no evidence that there are any secreto-motor nerves to the breast.

Lymphatic Drainage. The lymphatic arrangements of the breast are perhaps the most clinically important part of its anatomy, owing to their function in the spread of malignant disease. Most of the lymph from the periacinar lymphatics and probably also from the overlying skin drains obliquely backwards to the fascial plexus which lies at the interface of the deep and superficial fascia behind the breast. This plexus may be likened to a lymphatic marsh on which the breast rests. It empties by large channels, which probably accompany arteries and veins, chiefly to four sets of lymph nodes, namely (1) the pectoral group, lying along the lateral thoracic artery, (2) the subscapular group, arranged round the subscapular artery, (3) the apical group, lying round the medial part of the axillary artery, and (4) the internal mammary group, which lies inside the chest around the



(From the author's article in Lewison's "Breast Cancer" by kind permission of Dr E Lewison and the Williams and Wilkins Company

FIG. 191. Anatomy of the breast and axilla, from a dissection by Dr J M Lancaster The subscapular vessels in the cadaver lay anterior to the nerve to Latissimus dorsi

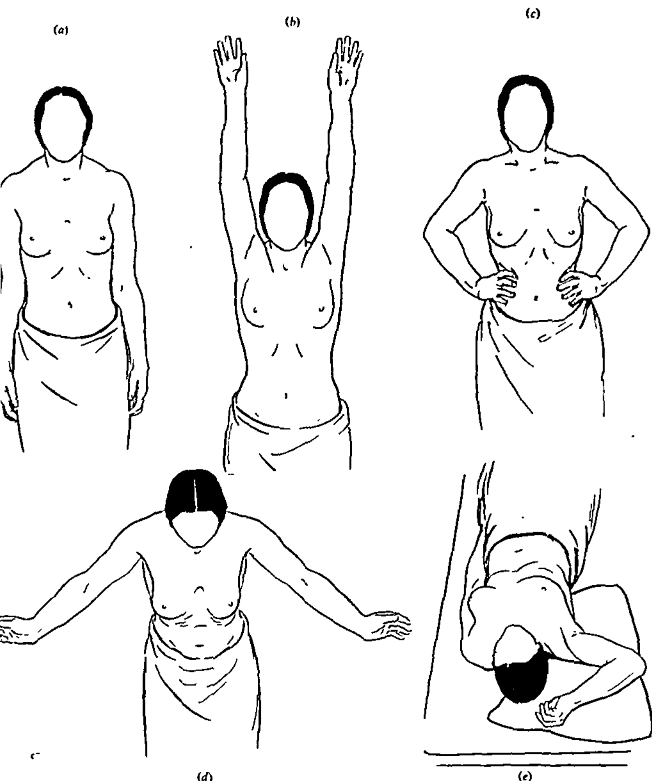


FIG 193. The clinical examination of the breast. (*a*) The patient stripped to the waist, stands before the surgeon, who may conveniently be seated. (*b*) The patient raises the arms above the head. (*c*) The patient places the hands on the hips and presses and relaxes as the surgeon directs (*d*) The patient bends forward, so that the breasts fall away from the body. (*e*) The patient lies flat with a pillow under the shoulder on the side to be palpated, and raises her arm above the head This manœuvre flattens the breast substance evenly over the chest wall for ease of palpation

internal mammary artery. The important drainage path to the apical axillary nodes is sometimes interrupted by the interpectoral lymph nodes of Rotter, which latter lie on the anterior surface of the costo-coracoid membrane. Other paths have been described but it is not thought that they are of much significance. They are (1) the direct path to the supraclavicular nodes, and (2) the path by which lymph might drain to the posterior intercostal nodes (which lie in the necks of the ribs) via the intercostal spaces.

The pectoral and subscapular lymph nodes are merely outlying parts of the axillary lymphatic system and discharge their lymph to the central and apical axillary nodes. The latter group discharges either directly into the great veins or into a node or nodes at the root of the neck whence lymph passes into the great veins. The only connection between the axillary and the supraclavicular nodes is that both often finally discharge into the same sentinel node near the great veins. The internal mammary lymph chain also discharges lymph at its upper end into the great veins, though a lymph node sometimes lies at its extreme upper end, behind the sternal head of the sternomastoid muscle, through which lymph passes on the way to the veins.

The circumareolar plexus of Sappey doubtless exists, but its function is probably to drain only the areola and nipple, and the two great trunks which appear in the older textbooks, draining it to the axilla, seem to be a figment of artefact or imagination.

PHYSIOLOGY OF THE BREAST

The breast, uninfluenced by nervous impulses, is controlled by a complex and ill-understood hormonal mechanism, which involves the ovary, the pituitary, the thyroid, the placenta, and the adrenal. Œstrogens cause hypertrophy of ducts. Progesterone has no influence unless acting in combination with œstrogens, when all the epithelial tissues of the breast grow. The formation of milk seems to depend on the action of the pituitary gonadotrophic hormone prolactin, acting in conjunction with adrenocorticotrophic hormone. A normal thyroid function is also required for lactation. The formation of milk is one thing, its discharge from the breast another. The latter function is brought about in part by the infant suckling, but it is also believed that certain cells in the alveoli, the myo-epithelium, have the power of contractility and are stimulated to contract by oxytocin, liberated from the pituitary by afferent stimuli reaching the hypothalamus. Suckling provides these afferent stimuli.

It is unfortunate that so little is understood of the hormonal control of the breast, because the breast is in a constant state of flux between puberty and the menopause and full understanding of its normal regulation would shed much light on its pathology.

CLINICAL EXAMINATION OF THE BREAST

The breast is usually examined badly. Though the routine to be described is not necessary in all its details in many cases, it will often bring to light points which would otherwise have escaped observation.

History. The history in a case of breast disease is seldom diagnostic, and it is on the results of examination that a diagnosis is most often made. The age of the patient, the presence or absence of pain, and the presence or absence of a lump are the points of greatest importance. The size and age of the patient's family (if she has one), troubles with lactation, her menstrual history, previous disease of the breast, any nipple discharge and the general health are points of interest and occasionally of importance.

Transillumination is not a method of much value. Its interpretation is most difficult in just those cases where it might be most useful, namely, when there is doubt whether or not a small lump is a cyst. The needle is not a diagnostic weapon. It should never be used to decide whether a lump is cystic or solid. This is not to belittle its value in therapy when a definite diagnosis of cyst has been made by other means.

CLASSIFICATION OF DISEASES OF THE BREAST

Diseases of the breast, like diseases in other organs, can be divided into broad categories. Though no classification can avoid criticism, the following scheme is useful and forms the basis for this discussion of mammary diseases. It omits very rare conditions.

A. CONGENITAL CONDITIONS.
- (i) Supernumerary nipples and breasts.
- (ii) Hypertrophy.
- (iii) Asymmetry.
- (iv) Atrophy.

B. TRAUMATIC CONDITIONS.
- (i) Hæmatoma.
- (ii) Fat necrosis.

C. INFLAMMATORY CONDITIONS.
- (i) Acute mastitis and abscess.
- (ii) True chronic mastitis (*a*) Non-specific.
  - (*b*) Tuberculous.
  - (*c*) Syphilitic.

D. DISORDERS OF METABOLISM (Hormonal disturbances).
- (i) Neonatal, puberty, and senile enlargement in males.
- (ii) Fibroadenosis.
- (iii) Mastodynia.
- (iv) Cystic disease (*a*) Galactocœle.
  - (*b*) Involutionary cysts.
- (v) Plasma cell mastitis, duct stagnation.

E. NEOPLASTIC CONDITIONS.
- (i) Benign (*a*) Fibroadenoma, pericanalicular, intracanalicular, cystadenoma.
  - (*b*) Papilloma.
  - (*c*) Lipoma.
- (ii) Malignant (*a*) Carcinoma.
  - (*b*) Paget's disease.
  - (*c*) Sarcoma.

CONGENITAL DISEASES

Supernumerary Nipples and Breasts. Supernumerary nipples are common, especially in men, and are seen on the lower chest and upper abdomen most often. They are of no importance, but can be excised if they are a cause of annoyance. Supernumerary breasts are very uncommon. They occur most often in the axillary area and lactate if their possessor should become pregnant. They should be excised.

Examination. In the clinical examination it is observation which is most often neglected; it may however be more important than palpation in arriving at a diagnosis. To start the clinical examination, the patient should strip to the waist and stand in a good light which falls squarely from the front ; the surgeon sits facing her, his eyes at breast level. The contour of the breasts and the nipple levels should be compared. The patient then raises her arms slowly from the side until they are straight above her head. Unsuspected lumps and skin dimples may thus be seen. She should then place her hands on her hips, alternatively pressing and relaxing as the surgeon commands. The appearance of skin dimples or abnormal movements of the nipple may be revealed. The patient next bends forward from the hips so that the breasts fall away from the body. This test may reveal changes in contour or a failure of one nipple to fall away from the chest as far as it should, a certain indication of abnormal fibrosis behind the nipple. The nipple is then inspected more minutely for tilting of its axis or retraction of a duct opening.

Palpation is the next step. It should be done with the patient lying. If the breast is of any size, a small pillow placed under the scapula on the side to be examined will tilt the patient's body, so that her breast rests evenly on the chest wall and does not fall to the side of her body, in which position its proper examination is impossible. It is also helpful to ask the patient to put her arm above her head. Palpation should always be gentle and warm examining hands are not only more comfortable for the patient but more sensitive. The breast is best examined with two hands, both with stroking movements and by using alternative pressure. It is in fact the fronts of the fingers, from the proximal interphalangeal joints to the pulp, and not the flat of the hands, which are most useful in palpation. The palpation should be systematic, quadrant by quadrant. The region of the nipple requires special care. In many patients the area under the nipple is softer than the rest of the breast and a circular hollow can be defined about three quarters of an inch in diameter, bounded by the firmer secreting tissue. Any filling or firmer consistency in this hollow area may indicate the presence of that bugbear of clinical diagnosis, a central carcinoma.

When a lump is found, the usual points which apply to a lump in any part of the body must be established as accurately as possible, namely whether it is solid or fluid, its outline, attachments, and mobility in the tissues. It is often not possible to determine these points with any accuracy if there is a small lump in a bulky breast. It is important to determine whether a lump discovered is attached to the skin, and this, if it has not already been seen while the patient was standing, may sometimes be determined by pinching up the skin and by pushing it to and fro in the region of the lump to discover if any localized diminution of movement can be discerned. Fixity to deep structures is determined by asking the patient to press her hands on her hips and thus to fix the pectoralis major muscle.

Both breasts should thus be examined. The axillæ are then palpated. This is often very difficult in fat women and it must be recognized that little useful information is gained in a clinically difficult axilla. The chief trouble in examining the axilla is to get the pectoralis major muscle to relax. The best way to do this is for a third person to hold the arm of the patient while she lies with the shoulder slightly abducted and slightly flexed. Without such assistance palpation from behind with the patient sitting up on the couch is as useful as any method. The examination concludes with palpation of the supraclavicular fossa and of the epigastric region.

Infections which arise through abrasions of the areola usually remain superficial. They resemble small boils in other parts of the body and they do not cause constitutional disturbance or even much pain. They are not infections of the breast tissue proper and their importance lies in their constituting a septic focus near the orifices of the breast ducts and in their potential interference with suckling. In the early stages they may be treated by a trial of penicillin, but the more successful and expeditious way in most cases is to evacuate the contained pus through a small incision, as far removed from the nipple as can conveniently be made, and to follow this by penicillin injections.

True acute mastitis in the lactating breast arises by infection, via the ducts, of an area of milk engorgement. Characteristically a sector shaped area of the breast becomes swollen, hot, tender, and painful and there is often considerable constitutional disturbance with fever and even rigors. With the passage of time, the pain becomes throbbing and the area of swelling smaller but more prominent and circumscribed. The skin is red and may become œdematous. In 3 or 4 days fluctuation may be apparent and in the untreated case, the abscess ultimately bursts through the skin. Sometimes the patient is less fortunate and the pus burrows and tracks in the breast substance forming loculi which, in extreme cases, may disorganize and deform the whole breast. It is said that such breasts are more liable to carcinoma in later years than the normal. Large loculated abscesses are certainly a nuisance at the time at which they occur, because fresh abscesses are very likely to appear in different areas and repeated incisions are needed. Abscesses may burst posteriorly and form a retro-mammary collection of pus on which the breast floats. A rare cause of retro-mammary abscess is due, not to mammary suppuration at all, but to bursting of an empyema through the chest wall.

Treatment. The factor of supreme importance in the early stages of acute intra-mammary infection is to relieve milk engorgement and to institute penicillin therapy, to a daily dosage of 1 million units. This will usually prevent abscess formation. Gunther has advanced the interesting idea that, in fact, acute mastitis in its early stages is purely an engorgement phenomenon and that the pain and the constitutional symptoms are due to extravasation of sterile milk into the breast tissue. Whether this is true or not, the relief of the engorgement at an early stage, either by suckling, by gentle but thorough manual expression or better by a modern breast pump, is the key to successful treatment. If an acute mastitis has been present for 3 or 4 days it has certainly become infected and it is very likely that pus will be present, even though fluctuation cannot be detected. Under such circumstances it is a mistake to attempt cure by the use of antibiotics alone. Antibiotics will not by themselves cure an established abscess: they may quench the flames but the fire continues to smoulder, and cases are sometimes seen where antibiotic therapy leaves a lump in the breast which will neither enlarge nor vanish and which gives much concern to both patient and surgeon for weeks. The correct treatment of an established inflammatory lump is the old-fashioned one of incision as soon as it is likely that pus is present. Fluctuation should not be awaited. If there is serious doubt as to whether pus is present, exploration of a breast lump with a large needle under general anæsthesia may be tried and an incision made if pus is encountered. If the inflammatory area is thought not to contain pus, dry heat is applied, if possible by diathermy or infra-red lamp, but otherwise by kaolin poultices backed with a hot-water bottle. Hot boracic fomentations, by making the skin of the whole area soggy, do more harm than good. Only when an incision has been made is chemotherapy used.

Hypertrophy, Asymmetry, Atrophy. Hypertrophy of the virginal breast, without general obesity, in girls between the ages of 12 and 20 is a very distressing condition. Fortunately it is not common. The breasts may attain such size that not only are they a cosmetic embarrassment, but they interfere with sleep. If the condition has been present for some time without showing signs of regressing when the periods are regularly established, plastic rectification should be advised. If no skilled plastic surgeon is available, it is better to do a simple mastectomy than to deform the chest wall with maladroit attempts to conserve the nipple. It is also extremely difficult for the amateur in plastic mammary procedures to achieve the necessary symmetry.

The breasts are commonly not exactly similar in size. This is no cause for alarm. A gross degree could be corrected in a young woman, who feared that her charms were thereby marred, by plastic procedures to the larger breast. Puzzling multiple rounded tumours are still occasionally encountered as a survival from the era of paraffin wax injections, the object of which was to enlarge the small breast. The tumours are masses of paraffin, surrounded by a foreign body giant cell reaction: they are harmless but unsightly.

Atrophy of the breasts occurs as a natural process in the old. It is occasionally seen in otherwise normal young women when it would perhaps more correctly be called a failure of development than an atrophy. It is most disfiguring when it is unilateral. *Prosthetic correction is the only treatment available.*

It is not proposed to consider abnormalities of the breast in hypogonadism, intersexuality, adrenal tumour, hypothalamic tumour or granulosa cell tumour. The changes are then but symptoms of a more serious general abnormality.

TRAUMATIC CONDITIONS

Trauma. Bruises and contusions of the breast are very common, but are usually small and resolve without trouble. Hæmatomata may result from blows. They are usually small and absorb unless they become infected—an uncommon cause of abscess. The treatment of definite wounds of the breast does not differ from the treatment of wounds elsewhere.

Fat Necrosis. Fat necrosis occurs in large breasts in women close to the menopause. Trauma is probably not its only cause. It is due to an area of fat being deprived of its blood supply. The dead fat is treated as a foreign body and provokes the typical reaction with foam cells, giant cells, and plasma cells, followed by fibrosis and sometimes calcification. The importance of fat necrosis lies in the exact mimicry of early carcinoma which it may give. It is a rare condition. The biopsy which removes the doubtful lump for microscopic examination also effects the cure.

INFLAMMATORY CONDITIONS

Acute Mastitis and Breast Abscess

The great majority of breast infections occur during lactation. They are caused by the staphylococcus aureus which gains access either through abrasions in the region of the nipple or along the ducts. The mode of entry determines, at least initially, the clinical pattern.

that the new forms of chemotherapy for tuberculosis may in future limit the surgeon's activities to aspiration or local excision of tuberculous areas. The rarity of the condition is likely to make the process of testing the new remedies protracted.

(c) Syphilitic Mastitis. Primary, secondary, and tertiary manifestations of syphilis are described in the breast. They must be extremely rare. My own experience contains a solitary instance of primary chancre. It is to be presumed that anti-syphilitic treatment should be tried if a case of breast syphilis is encountered and that any residual lump, such as might be left by a gumma, should be excised.

DISTURBANCES IN HORMONAL CONTROL

Hypertrophy in the Male

The breasts of boys commonly undergo an enlargement at puberty. The appearance of the tender discoid swelling beneath the nipple, perhaps 1 in. in diameter, is a source of anxiety to parents. It subsides within a few months in the great majority of cases, and it is only necessary to avoid chafe and blows, both of which cause pain. A similar hormonal change is also seen in new-born infants of both sexes. It rapidly subsides. Occasionally in adult men a similar change is seen, sometimes around the age of 60, when it may be presumed to be due to the male climacteric; or more commonly in younger men when trauma plays some part. In the latter, particularly if they are soldiers and have to wear web-equipment, excision of the area with preservation of the nipple and areola is the quickest means of getting such patients back to duty. In civilian life, a protective soft dressing and avoidance of chafe by braces should be tried before an operation is advised. The same clinical appearances are met with in men who are receiving stilbœstrol for prostatic carcinoma, with the addition of pigmentation of the areola.

Gynæcomastia. Gynæcomastia is a true hypertrophy of the male breast which assumes the appearance of the female. It may be associated with testicular tumours or testicular insufficiency from any cause, or it may occur in men otherwise apparently normal. The hypertrophy is sometimes unilateral. Obscure derangements of adrenal function are thought to be responsible for gynæcomastia. If a cause can be found it should be treated. Administration of testosterone may be useful, but if it does not assist, the breast tissue should be excised with preservation of the nipple.

Mastodynia and Fibroadenosis

The physiological changes which the female breast undergoes during the menstrual cycle may produce symptoms when such changes are more marked than usual, or when they occur in unduly sensitive people. Many women experience vague discomfort in the breasts shortly before the onset of menstruation, but occasionally it is so accentuated as to amount to pain. Moreover the pain may never completely disappear, though its sharpest exacerbations are before the onset of periods. Pain may be accompanied by a granularity or even nodularity of the breasts. It was to this condition that the term chronic mastitis was applied. It seems doubtful, however, whether chronic mastitis has a proper pathological standing. Pain may occur in an apparently normal breast. Nodularity may be found in a breast which has given no symptoms at all and is always present when the breast is preparing to lactate. All gradations may be discovered both in painful sensations and in degree of nodularity and these latter are not necessarily associated with

The technique of incision is briefly considered on page 393.

In early cases of acute lactational mastitis it is not necessary to interrupt feeding the child from the affected breast, unless pain makes it impossible; chemotherapy sterilizes the milk and the small concentration of the drug in the milk has no deleterious effect on the child. When a definite superficial abscess is present, pain will often prevent suckling, but it is not necessary to stop lactation. The affected breast should be emptied with the breast pump or by manual expression and, as soon as the pain occasioned by the abscess has subsided sufficiently, it should again be used for suckling. Lactation need only be stopped if there is serious intramammary suppuration. The older remedies of firm support, limited fluid intake, belladonna plasters, and salts by mouth, may be tried, but stilbœstrol (10 mgm. daily in divided doses for 3 days, "tailing off" for a further 6 days), is much more certain to arrest milk secretion.

The best treatment for acute mastitis is to prevent it. Ante-natal training in the care of the nipples, thorough emptying of the breast in the post-natal period and a well nourished and adequately rested mother are the cardinal points in prevention.

True Chronic Mastitis

(*a*) **Non-specific Mastitis.** Occasionally a chronic infection may supervene on an acute one and an area of induration which flares up acutely at intervals is the clinical indication that true chronic mastitis is established. More often chronic infection arises independently of lactation in patients with retracted nipples, which form a pit, difficult to keep clean, in which organisms can flourish. The latter track thence along the ducts. If a simple incision during an acute episode, followed by a long course of chemotherapy, fails to eradicate the infection, and cleaning of the nipple pit with spirit on cotton wool does not prevent fresh abscesses, it is justifiable to excise the affected segment of the breast. Even this is by no means always successful. Atkins has suggested that recurrent suppuration of this type is a chronic fistula running from the skin to an infected and blocked lactiferous duct and akin to a fistula-in-ano; he advocates passing a probe down the fistula into the duct and out again through the nipple. The track is then laid open and allowed to heal by granulation, in exactly the same manner as a fistula-in-ano.

(*b*) **Tuberculous Mastitis.** The breast is very resistant to the tubercle bacillus, and tuberculous disease is not common. The majority of examples arise from caseous material tracking into the breast from neighbouring structures, often within the chest, from infected anterior mediastinal lymph nodes, ribs, or pleura. Tuberculous axillary lymph nodes are another focus for this extra-mammary invasion of the breast by tuberculosis. It is likely that tuberculosis occasionally arises as a primary new infection through the nipple by contact with infected hands or linen. Blood spread to the breast is very rare.

Tuberculosis of the breast is most often seen as an abscess. Occasionally dense fibrosis is produced round a focus and may cause a lump which resembles a carcinoma; a history of tuberculosis should put the clinician on guard when he is examining a breast lump. If a lesion has been diagnosed as a tuberculous abscess, useful information can be gained by aspirating it, injecting lipiodol and seeing by X-ray where the lipiodol tracks. If clear evidence is obtained that the breast lesion is secondary to a focus within the chest, the latter clearly takes priority in treatment. It is useless to excise a breast or a portion of it if the track, through which infection reached the breast, is left running into the chest. In the past mastectomy has been advised for tuberculosis of the breast, but it seems likely

Cysts of the Breast

Cysts in the breast are of three varieties, by far the most common being the degenerative cyst formation found in so called "cystic mastitis." Galactocœle is rare and cysts which contain tumours are a separate entity which is considered in the section on duct papilloma.

(*a*) **Galactocœle.** The galactocœle occurs in the lactating breast or towards the end of pregnancy. It is due to obstruction of a duct, usually by fibrosis, from previous infection. Fibroadenomata, by causing pressure on a duct, are said occasionally to give rise to it in the lactating breast. The cyst, as its name implies, contains milk which may become inspissated and cheese-like; it is lined by necrotic tissue and encapsulated by fibrous tissue. It may be aspirated but usually recurs and should then be excised.

(*b*) **Cystic Disease of the Breast.** This condition, called cystic mastitis and known by a variety of other synonyms, is responsible for the great majority of breast cysts. Palpable cysts are more often single than multiple and they occur in the involuting breast, most commonly between the ages of 35 and 45. There would seem to be no good reason for sub-dividing cysts into various groups nor for regarding their cause as anything beyond obstruction of ducts. Their distribution in the breast is much the same as that of carcinoma, the upper outer quadrant being the favourite site. The cysts are thin-walled and tend to project forward from the breast tissue. Their contents are cloudy or turbid fluid. They are sometimes found in small clusters consisting of many small cysts, when their diagnosis is more difficult. Although they occur in much the same age group as does carcinoma, it is often easy to distinguish them by their regular contour, their mobility and the fact that most of the larger ones give obvious fluctuation. Transillumination, while it may help with very large cysts, is not as useful as one might suppose.

Treatment of a swelling, diagnosed as a certain cyst after clinical examination, is by aspiration and about two-thirds of them will not refill. The needle is not, however, to be used as a diagnostic weapon for elucidating the nature of doubtful swellings—it is a therapeutic weapon for treatment, and where there is doubt whether a lump is a cyst or not it should be excised. If, after aspiration, a cyst refills rapidly, if the aspirated fluid is blood-stained, or if the aspiration fails to abolish the lump completely, the area should be surgically explored. A breast which has produced one cyst may produce others at intervals and each time the swelling appears it should be viewed as a new problem and treated on its merits.

Cystic breasts show a slightly higher carcinoma rate than occurs in normal breasts and it is proper to do a simple mastectomy for a breast which is a mass of cysts.

Duct Stagnation and Plasma Cell Mastitis

In the ageing breast, the terminal parts of the main ducts often dilate considerably. If such a breast is examined in the post-mortem room, these ducts are found to contain creamy greenish material. Fibrosis around the dilated ducts may cause slow retraction of the nipple, and other clinical signs of this process of duct stagnation, or mammary duct ectasia, are the presence of a nipple discharge and the occasional palpation of distended ducts beneath the areola to give the so-called "varicocœle tumour" of the nipple. Sometimes the dilated ducts rupture and their contents then produce a violent inflammatory reaction in the tissues, with plasma and giant cell reaction, which slowly fibroses.

each other. It is thus impossible to define where the normal ceases and the abnormal begins, and there is no satisfactory correlation between clinical findings and symptoms. It is preferable to use the term mastodynia for the painful breast and fibroadenosis for the nodular breast, the latter term, unlike "chronic mastitis," having no associations with outworn theories and hypothetical conjectures.

Fibroadenosis. Fibroadenosis is, as its name implies, due to what is considered to be an abnormal degree of fibrosis, a process natural to the ageing breast, or to an abnormal degree of epithelial proliferation, a norm for which it is particularly difficult to establish in an organ whose physiology demands periodic waxing and waning of epithelial activity. Though histological studies show that the granularity of fibroadenosis is usually due to fibrosis, they sometimes reveal an overgrowth of the glandular tissue of the breast, which is so excessive that the term proliferative mastitis is applied. This proliferation may take the form of branching outgrowths of the duct systems to form new acini, a process called adenosis, or it may consist of a multiplication of the epithelial cells lining existing ducts. This latter process, termed epitheliosis, may fill the ducts with a solid cylinder of cells which are occasionally so atypical in appearance that the condition may reasonably be regarded as pre-cancerous. Nodularity due to epithelial proliferation is not distinguishable clinically from the much commoner form predominantly due to fibrosis. Epitheliosis is often seen in post-mortem specimens from breasts which have never given any trouble. The only way in which epitheliosis can be discovered is by biopsy and its infrequency and rather doubtful significance do not warrant biopsy on all cases of "chronic mastitis." It is only when a localized thickening gives rise to the sensation of a lump being present, that biopsy is indicated to elucidate its nature.

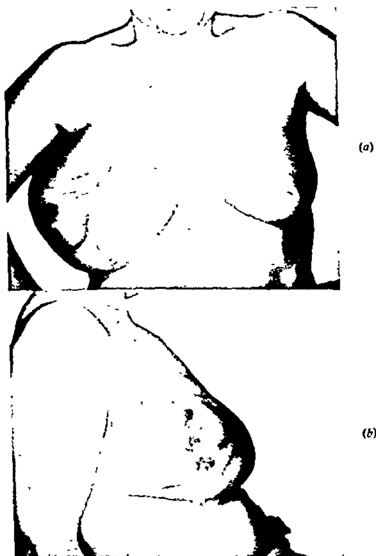
The clinical appearance of this elusive and ill-defined condition of fibroadenosis is the sensation of nodularity which the surgeon feels when he palpates the breast. The nodularity is not uniform in its distribution, and localized areas may be sector shaped. The upper outer quadrant of the breasts is predominantly affected. There are frequently well marked differences in nodularity between the two breasts. Soft axillary lymph nodes are often palpable. Such findings would be present, at one time or another, in the majority of healthy women.

Fibroadenosis is sometimes accompanied by cysts, and the latter will be considered in due course. Cystic disease of the breast is a well-defined clinical entity, in contrast to the indefinite and shadowy picture which uncomplicated fibroadenosis presents.

The treatment of mastodynia is to give reassurance that there is nothing wrong with the breasts, to explain the fact that discomfort in the breast is very common and will cease probably at the menopause, and to see that a well supporting brassiere is worn The only drug which seems to exert a definite action on breast pain is testosterone, and it is dubious therapy to prescribe so potent a substance, the full effects of which we know so little, for a symptom. Mastectomy should never be contemplated. Patients who have undergone removal of the breast for pain return with pain in the scar, in the other breast, or in some other part of the body. No treatment is required for the granularity or nodularity in the breast, which, giving no symptoms, is accidentally discovered. It is only when biopsy of a doubtful lump in the breast has shown great but still benign epithelial activity, that a simple mastectomy may be contemplated and even here we cannot be sure that such an area is in reality the precursor of malignant disease.

sarcoma, a course of radiotherapy to the lymphatic drainage areas would seem to be a wise, though in view of the rarity of the condition, an untested precaution.

(*b*) *Duct Papilloma.* The larger ducts in the breast frequently show some epithelial thickening. This sometimes amounts to a condition of papillomatosis when numerous small projections of epithelial cells are seen. In the main ducts epithelial overgrowth is



(*By kind permission of Mr R. Vaughan Hudson*)

Fig. 194 (*a*) and (*b*) Example of giant fibro-adenoma of the breast.

sometimes such as to give rise to a fully fledged papilloma, projecting into the duct and macroscopically recognizable. Such papillomata are usually situated close to the nipple and are sometimes palpable on clinical examination. They may be multiple in one duct or more than one duct may contain a papilloma. The classical symptom to which they give rise is bleeding from the nipple, but the discharge does not always contain blood; it may be clear. The management of cases of duct papilloma is so closely related to the treatment of nipple discharge that it is necessary to consider at this point the whole question of nipple discharges which occur in the absence of a palpable lump.

The inflammatory reaction is named plasma cell mastitis. The clinical picture presented is of the sudden occurrence of a painful swelling, in a woman of 55 or over, in the region of the nipple. After the acute phase has passed, there may be a small lump left with sufficient surrounding fibrosis to mimic carcinoma. Plasma cell mastitis is uncommon, but should be recognized as a condition which so closely simulates carcinoma that a radical mastectomy may be done mistakenly in place of the indicated local excision of the lump. This mimicry may be carried further by the nipple retraction which is caused by excessive fibrosis round the large ducts.

Patients occasionally suffer from recurrent episodes of plasma cell mastitis which cause much inconvenience and alarm. The recognized treatment in the older woman in whom the condition occurs is simple mastectomy, but Haagensen has found that excision of the nipple is satisfactory. The outlying breast tissue in post-menopausal women has not in his experience led to trouble when it has been left behind, and the operation is less extensive and less mutilating than a mastectomy. It would be unwise to attempt this operation on a breast which might lactate.

NEOPLASTIC CONDITIONS

Benign Tumours of the Breast

(*a*) *Fibroadenoma.* The fibroadenoma is the commonest benign tumour of the breast. It is an interesting growth because, though a tumour in the clinical sense of the word, it seems likely that it is in reality a circumscribed area of hormonal disturbance, akin to the process seen in fibroadenosis. The characteristic histological appearances are sometimes seen adjoining and inseparable from fibroadenosis and the microscopic appearances then suggest that both fibroadenoma and fibroadenosis are merely variants of the same pathology.

There are two forms of fibroadenoma, the pericanalicular, a hard variety, and the intracanalicular, a soft type, but the incidence and appearances are much less clear cut than textbooks have hitherto suggested. Classically, pericanalicular fibroadenomata occur in young women between the ages of 15 and 30. They may however occur at much later ages. They are sharply circumscribed, firm, and extremely mobile in the breast substance, slipping about under the examining hand in a very characteristic fashion. They are seldom larger than one inch in diameter and they may be multiple; or the removal of one may be followed by the appearance of another elsewhere. Intracanalicular fibroadenomata are less common. They are softer than the pericanalicular variety but very mobile. They are stated to occur in older women between the ages of 35 and 50, but they may be seen in younger patients and one tumour may show areas of both types. The intracanalicular fibroadenoma may grow to great size and occasionally ulcerates through the skin to give rise to an appearance similar to an ulcerated carcinoma. There is some question whether this giant variety of intracanalicular fibroadenoma is in fact essentially the same entity as the smaller variety because it may occasionally undergo sarcomatous changes. The large variety may also show cyst formation into which cauliflower-like masses of the tumour project (cystadenoma).

The treatment of the smaller fibroadenomata is to excise them. This is easy as they shell out of their capsules. In the larger ulcerated varieties, simple mastectomy is the proper treatment. If subsequent examination of the large tumours shows areas of

cases, namely contrast mammography where radiographs are made after the injection of radio-opaque media into the nipple ducts; and the cytology of the discharge, determined from stained smears.

Nipple discharges arise not only from duct papillomata and papillomatosis, but also from proliferative mastitis, from duct stagnation and from carcinomata which, very occasionally, are impalpable and only thus reveal their presence.

The treatment of a duct papilloma is evident from the foregoing. If it can be localized, the whole sector of breast tissue drained by the duct in which it occurs should be excised. If it cannot be localized, its existence remains problematical until the histological report has been received and the case should be dealt with as described above.

(c) Lipomata. Lipomata sometimes occur in the breasts of fat and usually elderly women. They are discovered as soft circumscribed tumours and they do not usually attain great size. They can be diagnosed clinically by the well-known characteristics of the lipoma if the possibility of their presence is borne in mind.

CARCINOMA OF THE BREAST

Of all diseases of the breast, carcinoma is by far the most important and difficult. Its importance lies in the fact that it kills about 8,000 women every year in England and Wales, that it is the commonest of all breast tumours and that it is the commonest of all forms of malignant disease in women. Its difficulty lies in its insidious onset, its tortuous and ill understood methods of spread and its extreme variability in behaviour. Perhaps the most surprising thing about it is the uncertainty which so often attends its diagnosis, since it occurs in an organ which is as easily accessible to palpation as any in the body.

Causation. Many theories have been advanced to account for the occurrence of carcinoma in the breast. There is some hereditary factor in that it occurs more often in some families than others. There is often a history of trauma, but small injuries to the breast are very common and any injury which is followed by a lump is remembered, whereas injury which subsides is forgotten. Trauma is sometimes thought to be the cause of carcinoma because a trivial blow may finally burst the tense capsule of a growing carcinoma, which later then proceeds to enlarge more rapidly once it is free of nature's first attempts at fibrous encirclement. In mice an unidentified factor is present in the milk of certain individuals which will produce numerous examples of breast carcinomata in the suckled offspring. If suckling is not permitted, the incidence of breast carcinoma in the offspring is low. Whether any such mechanism obtains in human beings is uncertain, but attempts are being made to shed light on this problem. Other factors which are believed to favour the onset of malignant disease are interference with the natural term and rhythm of suckling, imbalance of the hormones which are continually modifying the activity of the breasts, and serious disorganization of breast tissue by infection during lactation. In sum, the cause of carcinoma is as obscure in the breast as it is in nearly all examples of malignancy in other organs. What does seem certain is that clinical "chronic mastitis" has no definite relation to the onset of breast cancer.

Incidence. Carcinoma of the breast occurs most often in women between the ages of 45 and 55, but no age from 25 years to advanced senility is immune. There is no clinically useful difference between those who have and have not borne children, though the unmarried are slightly more liable to carcinoma than the married.

Nipple Discharges. Discharges from the nipple may be clear, milky, dark brown, or green or they may contain blood. A discharge which is at one time clear may at another be blood-stained and it is important to test any discharge from the breast for blood and to examine its cytology on more than one occasion.

Discharges which are milky and which are shown to contain fat globules are not of pathological significance. Discharges in young women which remain clear or milky on repeated examination can safely be watched, as can discharges of coloured fluid which



(By kind permission of Mr. R Vaughan Hudson)

FIG 195 Macroscopic section of giant fibro-adenoma From the case illustrated clinically in Fig 194 The tumour measured 6 in by 4 in

fails to react to the benzidine test. In women over the age of 40 any discharge, which can be expressed by focal pressure exerted by the blunt end of a pencil pressed on a particular area of the breast, demands that a sector shaped area of breast tissue, which includes the point at which pressure yielded the discharge, shall be excised for histological section. Perhaps the greatest anxiety arises in those cases of clear or non-bloody discharge in patients over the age of 40 where no focus can be made to yield the discharge by pressure. In such patients it is justifiable to wait and see, because the discharge is probably due to duct stagnation, but frequent re-examination of the breast is necessary. Discharges which contain blood always indicate the need for surgical treatment. If the lobe from which the bloody discharge comes can be located, a sector excision should be done. Where no such localization is possible a simple mastectomy should be advised in those patients who are over 40, and distasteful as it must be to any surgeon, this is probably also the safest course for blood-stained discharges in women under the age of 40. It must be emphasized that chemical proof that blood is present should always be obtained, because some dark discharges appear blood-stained when in fact they are not. It is to be hoped that further work may show the value of two further investigations in these difficult

the process by which carcinoma grows in continuity along lymphatic vessels. The presence of solid columns of cells in the lymphatics gives rise to fibrosis. This latter process fails to keep pace with the advancing column of cells, though the rear of the column is annihilated by fibrous constriction. Permeation is thus a variant of direct infiltration and the advancing tentacles of the carcinoma would be in continuity with the primary tumour, had the cells linking the growing edge with its source not been obliterated by fibrosis. It seems likely that permeation is of greatest importance when the larger lymphatic trunks have been blocked by invasion of the lymph nodes into which they drain. It is the process which gives rise to the satellite skin nodules of the late carcinoma, to the appearances of cancer en cuirasse and probably also to the recurrences sometimes seen in the skin flaps after operation.

The lymph nodes of the axilla are the main exit for lymph from the breast and they are therefore the most frequent site in the body for metastasis. It is a sad fact that more than two-thirds of all breast carcinoma patients have axillary metastases when they are first seen by the surgeon. The axillary system of lymph nodes nevertheless constitute a complex and extensive filter by which the passage of carcinoma cells is held up for some time.

Until recently the axillary lymphatic pathway was generally considered as the only important one. It is now clear that the internal mammary lymphatic system also takes an appreciable, though smaller, share in draining lymph from the breast. While two-thirds of the "operable" cases of carcinoma will show lymph node deposits in the axilla, no less than one-third show deposits in the internal mammary lymph nodes. Though the exact role of this pathway in the spread of breast carcinoma is still a matter for controversy, it is certain that the internal mammary node system is much less complex and less complete a filter than is the axillary system. It lies in direct contact with the pleura and it also offers a pathway for direct retrograde spread to both sides of the diaphragm and the dome of the liver. The table of the results of biopsy of the internal mammary chain, done mostly through the second intercostal space, shows the incidence of internal mammary invasion in a series of 150 cases. It demonstrates that carcinomata in the centre of the breast metastasize most often to these nodes and that growths in the inner hemisphere do so far more frequently than those in the outer hemisphere.

TABLE of the Lymphatic Spread of Breast Cancer in Relation to Site of Primary Growth, by Quadrant Analysis of 150 cases

	Site in Breast by Quadrants					Total
	Upper Inner	Lower Inner	Central	Upper Outer	Lower Outer	
All nodes free	11	4	1	28	5	49
Axillary nodes only invaded	6	2	4	34	6	52
Internal mammary nodes only invaded	4	0	2	2	0	8
Both axillary and internal mammary nodes invaded	10	7	10	13	1	41
Total	31	13	17	77	12	150

It is possible that there are other lymphatic pathways open to carcinoma for escaping from the breast than the axillary and internal mammary routes. Lymphatics may

Site. The left breast is affected slightly more often than the right. By far the most frequent site in the breast for the occurrence of carcinoma is the upper outer quadrant, but no area is immune. The upper inner and lower outer quadrants and the central area behind the nipple show an approximately equal frequency and only in the lower inner quadrant is carcinoma rare.

Surgical Pathology of Carcinoma of the Breast

Morbid Anatomy. The primary tumour in carcinoma of the breast varies in its appearance according to the rate at which it has grown and the defensive fibrous reaction which it has excited. The terms "scirrhous" and "encephaloid," to signify hardness or softness of the growth, are of no practical use. The majority of carcinomata are hard and when cut across impart the characteristic grating sensation to the knife, aptly likened to the cutting of an unripe pear. The cut section, which becomes slightly concave, reveals the white and yellow streaked appearance against a grey or pink-grey background, which is characteristic. Though an experienced morbid anatomist can usually recognize a carcinoma with his unaided eye, it is not always possible to say whether or not a lump is malignant without the microscope.

Morbid Histology. The usual histological type of breast cancer is the spheroidal or polygonal cell carcinoma. This may show varying degrees of differentiation and when it contains well formed tubules, it is termed a spheroidal cell adeno-carcinoma. The more rapidly growing varieties show columns and clumps of irregular cells without any recognizable arrangement and with hyper-chromatic nuclei and mitotic figures. Occasionally colloid changes are seen in breast carcinomata. Squamous cell carcinomata also occur occasionally and probably arise from the terminal parts of the main ducts. For the histologist the most difficult carcinomata are those in which the ducts are filled with masses of rapidly growing cells which have not burst their bounds and started to invade the tissues. Intra-duct carcinoma is the term used for this type of growth, and, while an intra-duct carcinoma is a potential cancer, it is usually difficult to demonstrate the infiltration of tissues, which is the hall-mark of malignancy—a fact which makes the prognosis of intra-duct carcinoma good. Intra-duct carcinomas often extrude worm-like masses when cut across, a peculiarity which has given rise to the name of comedo carcinoma.

Spread of Breast Carcinoma

Like most malignant growths, carcinoma of the breast spreads by four routes, namely by direct infiltration, by the lymphatics, by the blood stream and across the cœlomic cavities.

Direct infiltration may lead in late cases to invasion of the skin and ulceration. The growth may also directly invade the pectoralis major muscle and even the chest wall in neglected cases. But direct infiltration is of no great practical importance because it is amenable to excision and is not the cause of death in these patients.

The lymphatic spread of carcinoma of the breast is the centre of the whole surgical problem. Carcinoma cells, when they have gained entry to a lymphatic vessel, spread in two ways, by embolism and by permeation. It seems likely that invasion of the lymph nodes which drain the breast occurs most often by lymphatic emboli of carcinoma cells. Permeation, first fully described by W. Sampson Handley, in 1906, is the name given to

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by the liver. In Warren and Witham's autopsy series, many of whom had been intensively treated, lungs and liver were invaded in no less than 60 per cent of those dying of carcinoma. The bones follow next in order of frequency and it is the regional bones which bear the brunt. The sternum, ribs, and thoracic spine are the usual site, but all regions of the spine are frequently involved and so are the heads of the humeri, the heads of the femora and the pelvis. The skull may be attacked but it is unusual to see bony metastases distal to the knee or elbow. The opposite breast, the skin, and the adrenal glands are also frequently attacked, though it would be difficult to be certain in the first whether the carcinoma was secondary or a fresh primary growth. The brain is sometimes the site of an apparently solitary and early deposit. The eye is occasionally invaded. It is interesting to observe that, in this list, the organs closest to the breast seem to suffer most often from secondary deposits and this is an argument in favour of the predominating role of the lymphatic system in the spread of breast carcinoma.

Clinical Appearances

History. By far the most frequent reason why a patient with a breast carcinoma consults her doctor is that she has found a lump in her breast. The lump is nearly always painless until it is noted, and thereafter intermittent pricking and aching are felt. It is very seldom that the lump is found because it hurts, though this does sometimes happen particularly when the lump has been bruised by a slight accidental blow. Discharge from the nipple or alterations of the nipple, for example retraction, are sometimes the first abnormality noted by the patient, especially with carcinomata which lie behind the areola. Occasionally lumps in the axilla, due to axillary lymph node metastasis from a small tumour in the axillary tail are discovered by a woman who has no obvious breast lesion. It is astonishing how long some women will suffer their symptoms in silence, whether through indifference or fear, and even now carcinomata are sometimes encountered which are obvious not only to the hand and eye, but also, through ulceration, to the nose. Patients will occasionally conceal an advanced lesion in the breast when consulting their doctor for a lump in the axilla, an enlarged supraclavicular gland, jaundice, or a pathological fracture.

In taking the history the general points to be elicited have already been considered. They are usually of little help in the diagnosis, but the menstrual and lactational history of the patient is of interest.

Clinical Findings. The careful examination of breast cases, as already detailed, is carried out principally to elicit possible signs of an early carcinoma, so that they may not be missed in patients who at first sight appear to present a benign lesion or a normal breast. T[illegible]
taught th[illegible]
The progress of the disease usually next produces signs which are due to fibrosis around the growth with consequent shortening of Cooper's ligaments and abnormal fixity of the skin to the lump. The earliest stage of incipient skin attachment is the appearance of a shallow skin dimple which appears only in certain positions, e.g. when the arms are held above the head. This dimple later becomes a fixed dimple obvious to the eye at the first glance. As the growth progresses firm skin fixity over an extending area becomes apparent and reddening of the skin finally appears shortly before ulceration of the growth occurs. Before very obvious skin fixity is seen, nipple changes are likely in those whose

accompany the lateral perforating arteries, though they have never been proved to have transmitted carcinoma cells direct to the intercostal spaces.

Blood spread. The veins are fairly frequently invaded by carcinoma of the breast and undoubtedly blood-borne spread of carcinoma occurs. It is usually, however, a late phenomenon because carcinoma emboli in the blood stream find considerable difficulty in establishing themselves. They tend to provoke fibrin clots around themselves and to be walled off and strangled by fibrous tissue. Besides the normal system of veins which would bring any carcinoma emboli from the breast to the lungs, Batson has described in detail the role which the vertebral system of veins may play. In these vessels the blood may ebb and flow under the effects of varying abdominal and thoracic pressure and may allow the transmission of carcinoma emboli from the posterior abdominal wall to the brain without the mediation of the heart. These veins would be open to emboli from the breast via the intercostal spaces, but this would not affect the difficulty of carcinoma cells in establishing themselves via the blood stream. It may be concluded therefore that the blood stream is a potential path of spread but is usually of much inferior importance to the lymphatics in the *early* exit of carcinoma from the breast to the rest of the body.

Transcœlomic spread occurs only in the very late stages of carcinoma of the breast and is of less interest to the surgeon because he can do nothing curative for the patient who shows it. It is commonest as a transpleural manifestation. It also occasionally occurs in the peritoneum from an invaded liver and even, rarely, in the pericardium.

The spread of breast cancer depends not only on anatomical considerations, but also on the host-parasite relationship. Some tumours are highly "virulent" and grow rapidly. Some success has followed histological attempts to assess this "virulence" from the appearance of the cells, and grading of the malignancy of tumours bears some relation to the prognosis. This study has not yet reached a stage where it is of much help in estimating prognosis in individual cases. Besides the malignancy of the carcinoma, the progress of the metastatic process depends on the resistance which the host tissues show. Of this factor almost nothing is known except that recurrences of a breast carcinoma have been treated at intervals in a patient, without killing her, over a period of 42 years, a fact which seems to show that some sort of resistance to malignant spread does exist.

Local Reaction Around Carcinoma. The body does not accept the presence of an infiltrating neoplasm passively. As has already been said, we know almost nothing about resistance to the spread of tumours, but it is clearly obvious in every microscopic section that a fibrous tissue reaction is provoked by the presence of malignant disease. This varies greatly in its intensity but it is always present and it gives rise to many of the important clinical signs in early carcinomata, because fibrous tissue tends to contract. In the breast, in which the supporting stroma is closely connected by Cooper's ligaments with the skin; this contraction is susceptible of fairly early demonstration by the clinical tests already described.

Secondary Deposits in Carcinoma of the Breast

The lymph nodes draining the breast are the most frequent seat of metastasis, and [illegible] the axilla, closely followed by the nodes of the internal mammary chain. [illegible] until either the axilla or the internal mammary chain has transmitted car[illegible] and pleura are the most frequently [illegible] in advanced cases, after the regional nodes, and they are closely followed

A very misleading but definite clinical entity is the mobile circumscribed carcinoma. It is easy to mistake this type of carcinoma for a cyst or even a fibroadenoma, though it is never so mobile or so regular as the benign lesions. It is this clinical type which so often leads to tragedy unless the rule is strictly observed that all lumps in the breast must be excised unless their obviously cystic character indicates needle aspiration.

It is sometimes exceedingly difficult to decide whether or not a lump is present in the breast. The nodularity of fibroadenosis produces the most frequent puzzles, but comparison with the rest of the breast and with the opposite side will often assist greatly. The greater the experience of the surgeon in breast examination the easier it becomes to decide what is and what is not a lump. Nevertheless, even the most experienced are sometimes in doubt whether they feel a lump or an area of granularity and the only safe rule in such circumstances is to excise the doubtful area.

The inflammatory carcinoma, which occurs most frequently but not invariably during lactation, is another pitfall. The whole breast, or a large section of it, becomes painful, red, and tender and the appearance is mistaken for an acute mastitis. Here again an early biopsy is the only safe diagnostic rule in cases of "acute mastitis" which fail to respond to treatment within a matter of days.

The manifestation of Paget's disease of the nipple are considered in a later section.

In completing the clinical examination it must not be forgotten that both breasts, both axillæ, both supraclavicular fossæ and the epigastrium must be examined in all cases and that a radiograph of the chest is a matter of invariable routine.

Biopsy. The more experienced the surgeon, the more often does he resort to biopsy to prove the nature of lumps of which he is uncertain. The performance of a biopsy on the breast is never an occasion for shame or a proof of lack of clinical acumen. The better educated the public grows in the importance of breast lumps, the more necessary does biopsy become, because the smaller and more doubtful will be the lumps presented to the doctor. Though often the clinical appearances are all too obvious without pathological confirmation, only a stupid man will fail to recognize that the microscope is more accurate than his own unaided senses in early diagnosis.

In doing a biopsy, a general anæsthetic is necessary. Infiltrations with novocaine in the area of a malignant lump are obviously foolish. The least disturbance to a malignant growth is caused by incising it. This should be done if the lump is small and a small piece removed from the middle, followed by excision if it proves to be benign. A large [illegible] benign or not from the naked eye appearance of the cut section, but the microscopic examination occasionally yields a surprise. When biopsies are done, it is a great convenience to have a frozen [illegible] delay of [illegible] hours between a biopsy and full surgery does not appear however to influence the prognosis unfavourably, though the matter is difficult to assess*.

Staging of Breast Carcinoma

It is necessary to try to form some estimate of the stage which a breast carcinoma has reached for two important reasons. In the first place staging must be done, if not

* The paper by Nohrmann, quoted in the references, considers this problem.

carcinomata are not too far away from the nipple, and in large breasts with growths deeply situated near the nipple, the nipple is usually abnormally tilted towards the growth before skin signs appear. On raising the arms the nipple of the affected side rises higher than that on the normal side. These changes progress to gross deformity of the nipple. *Central carcinomata may produce only nipple signs, and uniform retraction of*



FIG 196 Advanced carcinoma of the breast, showing a lump, peau d'orange and retraction of the nipple.

the nipple in particular is their hall-mark. Deep fixity of a growth to the pectoralis major is usually a later manifestation than skin and nipple signs. Tethering of a growth to the pectoralis major may be tested by trying to move the lump when the pectoralis major is contracted. When there is fixity of the growth to the chest wall, it naturally cannot be moved over the chest even when the pectoralis major is relaxed. The sign of peau d'orange is always a late occurrence in breast carcinoma. It results from œdema of the skin, due to blockage of skin lymphatics, the deeper attachments of the hair follicles preventing uniform swelling of the whole surface and giving thus the characteristic "pig skin" appearance. When peau d'orange has appeared, other signs are so advanced that the nature of the underlying pathology is in no doubt.

(3) The presence of satellite nodules of growth in the adjacent skin.
(4) The presence of parasternal or intercostal subcutaneous nodules of growth.
(5) In the presence of œdema of the arm.
(6) When supraclavicular deposits are present.
(7) The presence of inflammatory types of carcinoma.
(8) When distant metastases are demonstrated.
(9) When any two or more of the following signs of locally advanced growth are present:

(*a*) Skin ulceration.
(*b*) Limited œdema of breast skin.
(*c*) Fixation of the tumour to the chest wall.
(*d*) Axillary nodes 2·5 cm. or more in diameter.
(*e*) Fixation of axillary lymph nodes to the skin or to the axillary contents.

Macdonald, Haagensen, and Stout have recently revised these criteria by deleting the carcinomata of pregnancy and lactation from their list of contraindications. They have added the provisos that, when there is clinical evidence of invasion of the axilla, a dissection of the supraclavicular triangle and the upper intercostal spaces should precede radical mastectomy and, if microscopic evidence of metastases in these situations is found, the case should be considered inoperable.

To many eyes the unrevised criteria gave too much latitude to the surgeon. For example, the presence of numbers of invaded axillary nodes or nodes stretching beyond palpation up into the axilla should be considered by themselves a contraindication to surgery as should fixity of axillary nodes to the skin or axillary contents. Ulceration of the skin over a growth is not by itself a contraindication to radical operation because it may occur, especially in very old women, before lymph node metastasis. Fixity of a primary tumour to the chest wall would by itself be held to contraindicate operation in many clinics.

In the majority of cases of carcinoma of the breast in clinical stage one, where in fact there is no evidence of spread beyond the breast, the treatment should be by radical mastectomy. Were it certain that the carcinoma was confined to the breast, a simple mastectomy would suffice, but it is unfortunate that many apparent stage one cases show axillary node invasion to the histologist; and it is to cope with this situation that radical mastectomy should always be done Many surgeons always give either pre- or post-operative radiotherapy, though more often the latter, to all their cases, but it seems that a full course of therapy is a needless infliction on the patient if the pathological examination of the post-operative specimen fails to demonstrate deposits in the axillary lymph nodes. A full [illegible] readily show. It does however seem wise always to give a localized application of radiotherapy to the area of the internal mammary chain, if this has not been surgically investigated (as it will not be in the great majority of cases).

It is in clinical stage two (where enlarged axillary lymph nodes are palpable but in which the axilla is not heavily involved and where there is no sign of more distant metastases) that there is a considerable cleavage of opinion about treatment. Radical mastectomy is nevertheless probably the treatment of choice, because it is possible that

formally, at least as a subconscious mental process, to decide on the correct treatment of the patient and for this purpose it must be done on the clinical findings. Secondly staging must be done in retrospect when the pathologist has examined the operative specimen, partly in order to plan further treatment and partly to arrive at some sort of more scientific conclusion in order to compare the efficacy of differing forms of treatment.

It is unfortunate that there are at least five different methods of staging, but this confusion is perhaps less unfortunate than it would otherwise be if any of the methods paid attention to the state of the internal mammary chain. For clinical staging this point cannot of course be ascertained, but for pathological staging it is a point of the greatest interest and importance and, without it, no classification is of much accuracy.

Both for clinical and pathological staging it is perhaps best to be simple and to stage breast carcinoma into three stages. Stage I comprises those cases where the growth is still within the breast, Stage II those where it has spread only to the immediate vicinity where it is still theoretically within an area which the surgeon can remove; and Stage III those where it has gone beyond the surgically accessible environs of the breast. In clinical staging one must remember also that widespread œdema of the breast, even in the apparent absence of axillary deposits, indicates advanced disease and that fixity either of the growth or of the axillary deposits to the chest wall also indicates an advanced stage.

Treatment of Breast Carcinoma

For the past 50 years the great majority of those cases of breast carcinoma which have received treatment have been treated by some form of radical mastectomy. How great an advance this operation was when it was first described, can still be judged very easily by reading Halsted's early papers It has however been realized that the results of radical surgery are not nearly as good as they are in some other forms of carcinoma. While radical mastectomy still remains the sheet anchor of treatment, two main schools of thought have emerged in those centres which have particularly interested themselves in the problems of breast cancer. The first believes that the classical radical mastectomy operation is not radical enough. The second considers that surgery should be curtailed in favour of radiotherapy. Any discussion of treatment must be coloured by this sense that we have arrived at a crossroads and that there is no signpost to indicate the correct way. Only actual exploration of the routes available, for which time is needed, will give the answer.

With these provisos in mind, it is still true to say that there are only two potentially curative weapons at present known, namely surgery and radiotherapy.

For early cases the results of surgery are very good. Dissatisfaction with radical mastectomy has arisen because the operation has been used in cases where the disease has already spread beyond the limits of possible excision. The work of Haagensen and Stout has been particularly valuable in preventing operations on the inoperable, a process which not only fails to cure, but often actually stirs the disease into quickened activity and shortens the patient's life. Criteria of operability have long been recognized but Haagensen and Stout laid down, after a study of the end results of operation, contra-indications to radical mastectomy which are worth quoting. They considered carcinomata inoperable under the following headings:

(1) Carcinomata of pregnancy and lactation.

(2) Carcinomata showing extensive œdema of the breast skin.

radical procedure logical. Sufficient time has not yet elapsed for its potentialities to be known, and it must still be regarded as a somewhat experimental procedure.

The treatment of the breast carcinomata of pregnancy is a difficult subject, because no one surgeon sees a large enough number of these cases to form a personal judgement. It is the usual practice in Britain to terminate the pregnancy and then treat the patient by radiotherapy, thereafter removing any residual mass by strictly local surgery. This is not, however, a universal view and Adair has discussed a series of 130 cases treated by radical surgery. Of 102 patients treated 5 or more years previously, 45 had survived. Adair found that when the axilla was not invaded the prognosis was but little worse in pregnant women. When the axilla was invaded, the outlook was much worse than in the non-pregnant. He has advised strongly in favour of terminating the pregnancy, advice which may be modified towards the end of pregnancy with the object of obtaining a live child, and must sometimes be abandoned in the face of the patient's religious views. It may be concluded that occasionally and in exceptionally favourable circumstances, radical mastectomy has a place, but that radiotherapy is the sheet-anchor of treatment; the pregnancy, if it is in its first 6 months, should be terminated.

Carcinomata occurring during lactation are more often of the highly malignant inflammatory type than are those of pregnancy. Inflammatory carcinomata should not be operated on but should be treated by radiotherapy. If they occur during lactation, the flow of milk should be stopped.

There is still some speculation as to the proper course to adopt if a patient who has recently been treated for carcinoma of the breast becomes pregnant. It would seem wise, in such circumstances, to terminate the pregnancy as soon as possible, unless there are compelling reasons to justify the risk that the pregnancy will light up hidden metastases which have escaped destruction. Pregnancy, whether terminated or not, is probably dangerous, and there is some evidence to suggest that it is in the post-partum period, when the breast should be at maximum activity, that recurrent carcinoma grows most rapidly.

It is believed by some that removal of the ovaries in young women with breast carcinoma is an added safeguard, even when the case is suitable for radical surgery. In advanced and inoperable cases in young women, bilateral oopherectomy is to be advised, but it is my practice not to do this if the primary treatment gives reasonable hope of cure. The value of bilateral oopherectomy has not been so conclusively proved as to justify adding a premature menopause to the deformity of mastectomy.

Palliation of Breast Carcinoma

Where curative treatment has failed, there is still considerable scope for prolonging and rendering tolerable the patient's remaining days. Until recently, radiotherapy was the only useful measure for palliation of the very advanced or recurrent carcinoma and it still occupies a very important place. Localized lesions often respond to irradiation and in particular the pain of localized bone lesions may be dramatically relieved by it. Deposits in mastectomy flaps, lesions in the brain, and lesions in the eyes may all respond well. Radiotherapy is of little use for pulmonary metastases, though the instillation of radio-active gold solutions into the pleura or peritoneum is sometimes highly gratifying in relieving pain and effusion.

The greatest recent advance in palliation has come from inducing changes in the sex hormone balance. The oft-quoted work of Beatson very early in the present century

the clinician may have mistaken inflammatory enlargement of nodes for neoplastic, a mistake which occurs in one of every four patients: or because the metastases felt may still be confined to the axilla, in which case radical mastectomy will cure the patient; and because recurrence in the axilla after operation is very unusual and an axillary clearance is thus likely to spare the patient the agonies of massive brachial plexus invasion. Operation should be followed, in those patients who are shown by the pathologist to have invasion of axillary nodes, by a full course of radiotherapy. It is widely believed in America that such radiotherapy adds nothing to a patient's chances but an equal volume of opinion in Great Britain holds that it results in a small but significant addition to length of life and to 5 year salvage.

When the primary tumour has transgressed the limits of the breast and its immediate environment, either because obvious distant metastases are present or because the local conditions have transgressed the criteria of Haagensen and Stout, the patient must be regarded as in stage three of the disease. It then becomes certain that surgery cannot hope to remove the carcinoma together with its ramifications. In such patients, the main therapeutic weapon is radiotherapy, with surgery as an adjunct to excise by local measures lumps which are ulcerated or otherwise a nuisance. Radical surgery should not be contemplated in these patients. Even when radiotherapy has so diminished the local evidence of widespread growth that an inoperable case becomes apparently operable, it is still a mistake to embark on anything more than purely local surgery for residual lumps. More will be said later about the palliation of advanced disease.

McWhirter has lately shaken the predominantly surgical outlook on early carcinoma of the breast by showing that by combining quite a limited simple mastectomy with radiotherapy, he can obtain survival rates which compare with the best surgical results. His excellent results remain to be repeated by others. If they can be, and if it can be shown that his method of treatment does not sacrifice cure in the early cases for longer survival in the incurable, there is no doubt that radical mastectomy will become an operation of historic interest only. McWhirter's thesis, in brief, is that if a carcinoma is confined to the breast, only a simple mastectomy is needed, though radiotherapy must always be added in case the clinical appearances are deceptive: if the carcinoma has reached the axilla or the internal mammary lymphatic chain, it is incurable by surgery and radical mastectomy is a waste of time. McWhirter's work, besides offering a potential improvement in treatment, has also had the invaluable effect of shaking surgical thinking out of its complacency and compelling surgeons to reconsider their fondest conceptions. What McWhirter has not explained is why others using his technique have so far not repeated his results; nor has he demonstrated that the "healthy" looking carcinoma cells often to be seen in carcinomata and their deposits after a full course of irradiation are incapable of further growth.

The demonstration of the frequency with which the internal mammary lymph chain is invaded has not only strengthened McWhirter's arguments, but it has also stimulated surgeons to consider radical excision of the major part of the internal mammary chain by extending the scope of the classical radical mastectomy. This operation is technically feasible, in the form described by Urban, with very little additional danger or deformity to that of radical mastectomy, though with considerablly greater efforts for the surgeon. The realization that the internal mammary chain is a primary lymphatic drainage area of the breast, though a smaller one than the axillary system of nodes, makes the extended

seems to have been the first attempt to modify the hormonal environment of breast carcinomata and Beatson did this by surgical removal of the ovaries. Then, as now, not all tumours responded to changes in the circulating hormones and Beatson's many failures were remembered while his few successes were forgotten. In 1939 Loeser introduced the administration of testosterone to palliate advanced breast carcinoma and thereby re-awakened interest in the whole question of treating advanced breast growths by changing their hormonal environment.

No example of cure by hormonal modification has ever been claimed. The most successful way of using the sex hormones is to change their balance as far as possible from what it was when treatment started. Thus in women who have not passed their menopause by more than 5 years, testosterone is the drug of choice. The testosterone may be given by mouth, by injection, or by implantation of pellets of the hormone into the tissues. Testosterone propionate by injection, 150 mg. twice weekly, may be taken as an accepted method of administration, but there is much variation in the testosterone preparations used and in their dosage. Testosterone may entirely fail to produce any effect. It may cause dramatic shrinkage and disappearance of deposits. More commonly, it increases the patient's comfort and sense of well-being without markedly affecting the objective signs of malignant deposits. Not only does testosterone masculinize patients but it causes increase in weight by water retention and it may, in patients with damaged kidneys, precipitate renal failure. Perhaps its most striking results are achieved in some examples of bone deposits where not only is pain relieved, but recalcification of the bone takes place. The effectiveness is by no means always limited to those younger than 5 years past their menopause, and it may have striking results in very old women. Among its gratifying effects is the sense of well-being and cheerfulness which it often evokes.

In women well past their menopause, the œstrogens are more likely to palliate than the androgens. This is indeed the most successful of all forms of hormone therapy. Œstrogens are most usually given as stilbœstrol in dosages of up to 15 mg. daily by mouth, if the patient can tolerate such dosage without nausea or other side effects. Ethinylœstradiol has also been used in dosages of up to 3 mg. daily by mouth and is said to cause less nausea. The œstrogens are however more dangerous in that increased growth of carcinomatous deposits may sometimes be provoked and the experimental evidence that œstrogens favour the onset of breast carcinoma in animals is fully confirmed. It is said that œstrogens are more likely to affect favourably pulmonary deposits than are androgens.

The work of Huggins has recently introduced the operation of adrenalectomy to the field of palliative hormonal treatment. The adrenal is an undoubted source of sex hormones and the recent isolation of cortisone has allowed these glands to be ablated without killing the patient in an Addisonian crisis. Much the same partial success has attended adrenalectomy as with other forms of hormone therapy. It is now practised on patients for whom no other therapy remains untried. It is combined with bilateral oophorectomy and both ovaries and both adrenals may be removed at one time without undue risk. The operation itself presents no special difficulties but the pre- and post-operative managements are difficult and demanding. In about one case in every four there is a dramatic response which may sometimes last up to 3 years. On the whole it appears that the most favourable results are achieved in the slower growing and better histologically-differentiated tumours. Often the operation makes no difference to the patient's deposits.

It appears that complete removal of the pituitary gland may achieve effects similar to those of adrenalectomy, but there is as yet no reliable information on a sufficient number of patients to form any opinion on what seems a formidable undertaking. A less difficult manner of extirpating the pituitary than by operation has lately been desscribed by Forrest who destroys the gland by implanting radon seeds with an ingenious introducer.

It thus seems that we work very largely in the dark with all forms of hormone treatment. Some patients show a miraculous response—others none whatever. The response is always temporary and there seems no reason for giving hormone treatment as a safeguard to early cases who have been operated on, unless obvious signs of distant metastases appear. But when surgery and radiotherapy have failed to cure or relieve, it is always worth while to try the effects of hormones, and if one form fails, to attempt another.

Where everything has been tried without avail, it is worth considering, in patients with intolerable pain, whether the neurosurgeon can render aid by the interruption of the pain pathways, either by operation or the intrathecal injection of alcohol.

The Results of Treating Operable Breast Carcinoma

Length of Life. In considering the results of treatment, it must be remembered that the average length of life from the first symptom to death is about 3 years in patients who receive no treatment at all. This surprising figure cannot now be scientifically investigated, but has been reached from old records and notes of the pre-Listerian era. Many surgeons will recollect examples of survival for 7 or 8 years in cases of untreated carcinoma, particularly in older women. The futility of estimating the results of treatment in any period of less than 5 years from the inception of that treatment is obvious from these facts.

The operative mortality for radical mastectomy is very low. It should not exceed 1 per cent in large series and the deaths are usually due to such unavoidable catastrophes as pulmonary embolus or coronary thrombosis. Nor does the mortality for extending the radical operation to include the internal mammary chain appear to exceed that for the standard operation, though there is some selection of the fitter patients for the bigger operation.

The patient who, at radical mastectomy, is shown by the pathologist to have no lymph node deposits in the axilla, has a 70–75 per cent chance of living 5 years and a 45 per cent chance of living 10 years. Her actual chance of not dying from recurrent carcinoma is better than this by a figure which represents the normal expectancy of death for the population of her age, a factor which most papers do not take into account. It seems probable that patients whose internal mammary lymph nodes are free from invasion in addition to the nodes in the axilla, will show a 5 year survival of about 85 per cent.

A very sharp decline in the patients' chances of 5 year survival after radical mastectomy occurs when the axillary lymph nodes are invaded. It falls to about 35 per cent and there is a continuing mortality which brings the 10 year survival to under 20 per cent.

Comparable results from [illegible]
are d [illegible]
into [illegible] the state of the axilla and this is not certainly known unless the nodes are

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In the practical realm of diagnosis and treatment, a biopsy should be done on any case of eczema of the nipple which does not very quickly clear up from simple applications. If Paget cells are seen, a radical removal of the breast should be performed and the prognosis is then excellent if the axillary lymph nodes are free from tumour cells.

Sarcoma of the Breast

Sarcomata are extremely rare in the breast. They arise either from the fibrous tissue of the breast or in connection with fibroadenomata, and their diagnosis is often not made until a histological report is received. Sarcomata of the breast spread more often by the blood stream than by lymphatics and they are said to show a tendency to local recurrence on the chest wall. It would seem wise always to do a wide simple mastectomy for large fibroadenomata which have shown rapid recent increase in size and to follow this with a full course of radiotherapy if sarcomatous changes have occurred in the tumour.

Fibrosarcomata may occasionally occur in the subcutaneous tissues of the breast; they are not sarcomata of the breast proper.

NOTES ON OPERATIVE TECHNIQUE IN BREAST DISEASE

The following notes are not intended to supply full details of operations on the breast which more properly belong to textbooks of operative surgery. They are intended to comment on the underlying principles which should guide operations; and to explain what the surgeon attempts to do.

Most women regard their breasts as a part of their feminine quality and the loss or disfigurement of them is a blow to their "amour propre" which a man finds it difficult to understand. There are times when loss or disfigurement are inevitable and, particularly in the treatment of carcinoma, a patient's feelings must be sacrificed in the interests of her life. This does not absolve the surgeon from a sympathetic understanding of the psychological blow which the treatment involves and, within the limits demanded by effective treatment, he should try to plan his incisions and his procedures so that their effects may be as inconspicuous as possible.

Minor Operations on the Breast

In making incisions in the breast, it must be remembered that radial incisions in the breast substance are likely to do the least damage to the duct systems. But the skin incision need not be in the same line as the breast incision. Radial skin incisions in the upper inner quadrant are particularly objectionable from a cosmetic viewpoint because they may be visible above the neck line of a dress. A skin incision parallel with the neck line of the dress can here be undermined sufficiently to allow an ample radial incision in the breast substance.

In opening breast abscesses it is wise to make a small first incision, large enough only to introduce the finger for gentle exploration. A second small incision can then be more accurately placed to obtain dependent drainage if necessary, and the first incision loosely sutured. Rough digital manœuvres and the vigorous breaking down of loculi in an abscess should be avoided.

Lesions in the region of the nipple may be explored through a T incision, which gives a great deal of room for its size and is inconspicuous when it heals. The short limb of the T is peri-areolar, at the junction of areola and skin, the long limb being radial.

removed by radical operation. McWhirter has given his 5 year survival rate for "operable" cases—that is approximately those in stages one and two—as 55 per cent. Comparable surgical figures for stages one and two together show the same sort of results from radical mastectomy at, for example, the Memorial Hospital, New York.

There have not been wanting those who believe that treatment makes very little difference to the length of a patient's life. Others believe that the type of treatment administered, provided it is carefully done, makes little difference to results. With the infinite number of variables in the behaviour of tumours and in the method of treating them, it is impossible to get very close to the truth in general, and quite impossible to foresee in any particular individual what will happen—only a probable forecast can be attempted.

Other Benefits of Treatment. Length of life is not the only benefit of treatment. Quality of life may be even more important, though it is impossible to measure accurately. A well planned radical mastectomy in a properly selected case will nearly always spare a patient the disasters of local or axillary recurrence. Radiotherapy in appropriate cases will likewise achieve great benefit in relieving local manifestations such as ulceration.

Complications of Treatment. Complications of treatment occasionally arise and perhaps the most distressing is that of brawny arm. This gross form of œdema is nearly always encountered in fat women who have had septic interference with the healing of their operation wounds in the region of the axilla. Radiotherapy increases both the incidence and the severity of brawny arm. Some degree of swelling of the arm, not amounting to a brawny arm, and not interfering with the patient's use of the arm, is very common after radical mastectomy. The other sequel which is sometimes distressing is limitation of movement at the shoulder. This again is made worse by radiotherapy. It will often yield to manipulation under anæsthetic, and physiotherapy, unless it is due to a tight band of scar tissue. Plastic correction of the latter is sometimes worth attempting.

Paget's Disease of the Nipple

While simple eczema of the nipple may occur, it is never wise to make this diagnosis clinically because an apparently simple eczema often denotes the presence of an occult central carcinoma. This combination of nipple eczema and mammary carcinoma is called Paget's disease. Paget's disease occurs in two forms, as an apparently primary change where the rest of the breast seems normal; and in a secondary form when, in addition to the nipple eczema, there is an obvious carcinoma present. The secondary form naturally causes no difficulty in decisions on treatment. The apparently isolated eczema is however a very considerable stumbling block.

The characteristic feature of Paget's disease is the Paget cell, a large hydropic cell lying in the deeper layers of the epidermis. Its nature has never been finally settled. It has been contended that it is merely an œdematous change in squamous epithelium caused by lymphatic blockage from a hidden breast carcinoma, that it is a malignant change in squamous epithelium occurring simultaneously with a malignant breast change, and that it is a carcinoma cell of duct origin which has migrated to the epithelium. The latter view seems the most likely because the breast growths associated with Paget's disease are always central and originate from the larger ducts. Paget cells in the skin have also been described in the neighbourhood of other carcinomata, e.g. of the anal canal.

Radical Mastectomy

The methods of performing radical mastectomy are legion. Not only the skin incisions, but also the steps of the operation are the subject of countless variations. It is believed that the following short account of the operation would be acceptable to most surgeons.

Most of the time spent in a radical mastectomy is taken up with hæmostasis, and any

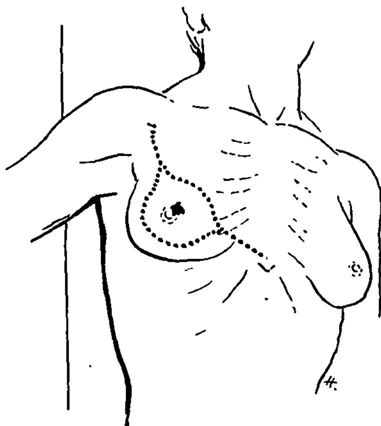


Fig 198 Incision for radical mastectomy. The tumour is indicated in black and lies at the centre of the skin to be removed Note that the upper limb is directed towards the coracoid process rather than on to the arm

measure which will diminish bleeding will save much time. Formerly the use of chloroform as an anæsthetic achieved this. It is doubtful whether modern hypotensive techniques are any safer than chloroform, but fortunately bleeding can be considerably diminished by posture. The patient is secured to the table so that she can be tipped, feet down, to a maximum angle of 30 degrees and variations in this angle will give a considerable and controllable variation in the blood pressure.

It is very convenient to be able to move the arm on the effected side during operation so that the axilla can be cleared. In positioning and towelling the patient, an arm board is used and covered with a sterile towel, bandaged on to it. The arm is also wrapped in a sterile towel so that the surgeon's assistant can handle it as directed. Two assistants are a great convenience in the operation.

When segments of the breast substance must be excised, they should be sector shaped and take the tissue to the edge of the breast, so that (in so far as possible) breast tissue is not left without duct connections to the nipple. The edges of the gap left in the breast substance are then coapted with buried interrupted sutures, introduced with a curved cutting needle. This minimizes the deformity of removal of a segment of the breast, and restores its circular disc-like appearance.

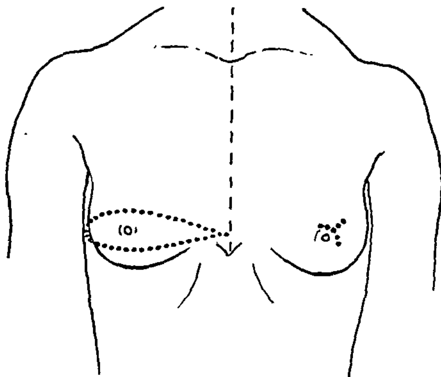


FIG 197 Incision for simple mastectomy on the right, and on the left, T incision for exploring lumps in the region of the nipple

The secret of successful healing in all breast operations, minor as well as major, is painstaking and meticulous hæmostasis. Large vessels should be tied, but the many little oozing points seen in, for example, a segmental resection, can be dealt with only by the coagulating diathermy. A perfectly dry wound does not require draining. Hæmostatic gauzes and sponge should not be used as they provoke a marked reaction in the breast.

Simple Mastectomy

The performance of a simple mastectomy demands wide undermining of the flaps if the operation is not in fact to be only a partial mastectomy. In so-called simple mastectomies, the axillary tail of Spence in particular is often left behind and has been known to become the seat of a carcinoma later. A simple mastectomy can be most neatly done through a transverse elliptical incision which removes the nipple and leaves a scar which is completely hidden by a brassiere. To do a complete simple mastectomy through a transverse scar demands a long incision beginning in front in the mid-line and ending postero-laterally at the posterior axillary line.

mind the problem of closing the wound until he reaches this stage, because he may otherwise be tempted to "cut his corners" in doing a proper ablation of the malignant process.

Once the skin flaps are cut, the limits of the excision are defined by cutting down on to the sternum medially, to the chest wall inferiorly and through the thickness of the pectoralis major above, between its sternal and clavicular heads. The upper end of the lateral flap is then retracted and the sternal head of pectoralis major sectioned as near its

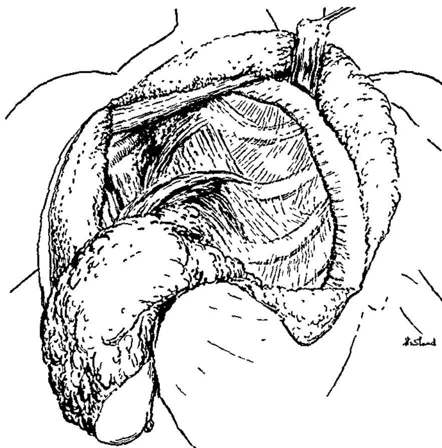


FIG. 200. Radical mastectomy. The pectoralis major has been divided between its sternal and clavicular heads above, and the sternal head sectioned, close to its origin medially. The pectoralis minor has been [illegible]
and the pectorali [illegible]
removal of the br[illegible]
dissection will be [illegible]

insertion into the humerus as possible. The upper part of the origin of the pectoralis major from the sternum and chest wall is now divided, to leave the costal cartilages and intercostal muscles exposed, and it is at this stage that the anterior perforating arteries are divided: they can usually be seen and ligated before they are cut if a little care is exercised. Retraction of the clavicular part of pectoralis major will now expose the origin of pectoralis minor which should be divided as close to the coracoid process as possible. In many patients the clavipectoral fascia can then be demonstrated by inserting a finger through the gap left by section of pectoralis minor and directing it medially and laterally, and this much facilitates accurate division of the fascia and the ligation of the

In making the skin incision it is necessary to remove enough skin and the incision around a growth should not be closer than 1½ in. from its edge. This means removing a circle of skin at least 4 in. in diameter, and often bigger. From this circle, linear prolongations will be needed to gain appropriate access, the lower prolongations being toward the xiphisternum and the upper towards the coracoid process. Incisions which travel on to the arm, though they give good access to the lateral end of the pectoralis major, cannot be easily concealed, and should not be made. The first incision should be

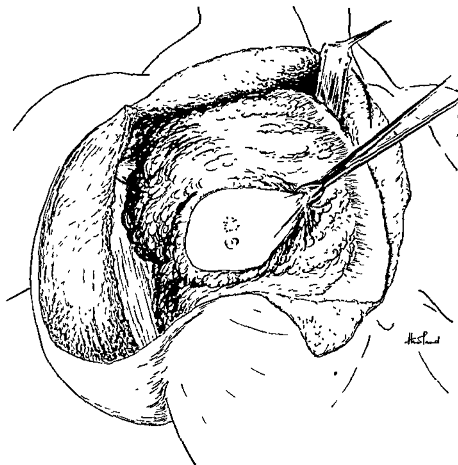


FIG. 199. Radical mastectomy Flaps are shown cut to the limits of the excision and at the four edges the incision has been deepened to the pectoralis major above, the latissimus dorsi laterally, the chest wall below, and the sternum medially

made carefully, so that it goes no deeper than the dermis, and bold incisions which go deeply into the fascia should be avoided. Thin skin flaps are then raised to the proposed limits of the excision, namely to the mid-line medially, to the anterior edge of latissimus dorsi laterally, to the groove between sternal and clavicular heads of pectoralis major above and to a level an inch below the obvious lower limit of the breast. The object of raising thin flaps is to remove all the breast substance and as much as possible of the lymphatic networks underlying the skin. A "thin flap" policy, together with removal of a good deal of skin, entails frequent grafting because suture of thin flaps under tension inevitably spells necrosis—a sufficient bugbear of thin flaps even without tension. It is an axiom that the surgeon operating for carcinoma of the breast must banish from his

Closure of the wound follows and may be particularly difficult when the carcinoma lay towards the axillary tail. It is very necessary to close the axilla with a flap and some rotation of the lateral flap may on occasion be necessary: it is achieved by an incision running posteriorly and then upwards on to the back. Defects in flap cover on the chest wall are covered by split skin grafting. The general surgeons might well take lessons from their plastic colleagues when it comes to closure of breast wounds.

The treatment of skin grafts in breast surgery owes much to the observation of D. H. Patey that pressure over a graft, particularly if it rests on a bed of muscle, is unnecessary. It is in any case impossible to maintain a good pressure dressing on the chest wall. All that a skin graft in this situation needs is protection from rubbing. After it has been spread on its bed, it requires only a shield of perforated zinc, held away from it by strips of thick orthopædic felt, which are glued to the skin surrounding the grafted defect with Portex adhesive. The surface of the graft can thus be kept dry and the percentage "take" is higher than with orthodox techniques.

Radical mastectomy wounds are particularly liable to show collections of fluid under the flaps. This fluid comes chiefly from the axilla, and the nuisance it causes can be almost entirely eliminated by suction drainage. A long rubber tube (½ in. external diameter), with side holes cut in its distal 6 in. is introduced, through a stab incision near the costal margin, where its entry is well away from the septic axillary skin. It is connected to a silent suction pump, maintaining a negative pressure of 2 or 3 cm. of water. The first 24 hours may yield 8–16 ounces of fluid and suction is usually maintained for 3 or 4 days. Not only does this keep the wound dry but it enables nearly all dressings to be dispensed with. It has the further inestimable advantage of making atmospheric pressure bring the flaps into close contact with the chest wall and reduces to a minimum the troublesome collections of fluid under the flaps which often mar convalescence.

Ablation of the sternal head of pectoralis major is a tenet of the classical radical mastectomy, but I doubt if it is in fact necessary. The key to the clearance of the axilla is the removal of the pectoralis minor. By elevation of the arm, the sternal head of pectoralis major can be sufficiently relaxed to permit of full exposure of the axilla to the outer border of the first rib. The advantages of leaving the sternal head are cosmetic and functional and the arm seems less liable to suffer from severe post-operative œdema. Recurrence in the pectoralis major, when it has been preserved in radical mastectomy, is not seen, and if (as is now generally believed) the early spread of breast carcinoma is by lymphatic embolus, it would be unlikely that an embolus of carcinoma cells in transit would be left behind in the muscle.

Extensions of Radical Mastectomy

The frequency with which the internal mammary chain is invaded in apparently operable cases of breast carcinoma has naturally led surgeons to ponder surgical attack on these nodes. The nature of this attack has varied from simple biopsy-like procedures to a resection of a segment of the chest wall with the underlying internal mammary vessels and nodes. The benefit of these operations is still unproved and their evaluation awaits the passage of time.

Biopsy of Internal Mammary Lymph Nodes. This is a simple manœuvre if taken slowly. The intercostal muscles are cut cautiously across their fibres midway between and parallel to adjoining costal cartilages, from the edge of the sternum to the lateral end of the

thoracoacromial vascular bundle and its branches before they are cut. With gauze and non-toothed dissecting forceps, fascia, and fat are pushed caudalwards to expose the axillary vein, small vessels being seen and double tied before they are cut and the pectoral nerves divided. The anterior and inferior surface of the axillary vein is thus left clean and exposed. In continuing the dissection in a plane posterior to the axillary vein, the nerve of Bell (nerve to serratus magnus) and the nerve to latissimus dorsi must be sought and the former, at least, preserved. The nerve of Bell lies in the fascia covering serratus

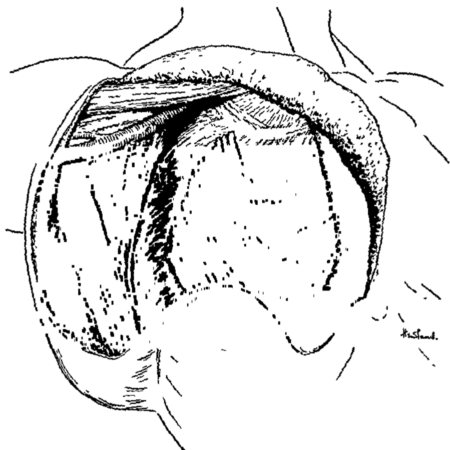


FIG. 201. Radical mastectomy. The wound after removal of the breast and prior to suture.

magnus, and may be pulled away from the chest wall if the fibres of the muscle are bared. A little patience with blunt dissection at this stage will reveal the nerve, though confusion is sometimes caused by the large lateral branch of the second intercostal nerve which emerges from the chest wall with its vessels; these latter must all be divided. In continuing the dissection outwards, the nerve to latissimus dorsi lies on the surface of the subscapularis muscle. If the presence of invaded nodes makes it difficult to preserve, it should be resected, as should the large subscapular vessels which lie immediately lateral to it; but all these structures may be preserved if lymph node metastasis does not complicate the dissection. Working from above and medially to downwards and laterally the chest wall is now cleared by sectioning the origins of pectoralis major and minor and leaving, clean and exposed, the chest wall, the upper part of the rectus sheath, the digitations of serratus magnus, the subscapularis, and the latissimus dorsi.

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Biopsy of Internal Mammary Lymph Nodes. This is a simple manœuvre if taken slowly. The intercostal muscles are cut cautiously across their fibres midway between and parallel to adjoining costal cartilages, from the edge of the sternum to the lateral end of the

cartilages. Blunt dissection then reveals the internal mammary blood vessels embedded in fibro-fatty tissue and lying just clear of the edge of the sternum. Lymph nodes may be picked out from either side of the vessels. The nodes here are often exceedingly small. The only serious hazard in the procedure is the danger of wounding the internal mammary artery—it is a vessel of large size. The pleura may be torn but positive pressure anæsthesia is the complete answer to this event. It is not possible to resuture the intercostal muscles at the conclusion of the exploration.

Excision of the Internal Mammary Lymphatic Chain. The entire internal mammary lymphatic chain could only be removed by operations of heroic magnitude because it runs up into the neck behind the clavicle. In practice the chain can fairly easily be excised from the level of the first to the sixth ribs and it is to this part of it that the breast lymphatics discharge. To perform this excision, it is necessary to resect a block of the chest wall, measuring approximately 4 in. by 1 in., and including the second, third, and fourth costal cartilages, the intervening intercostal muscle, and the subjacent internal mammary vessels and nodes, which adhere to the chest wall by reason of their perforating branches. The internal mammary vessels are ligated and divided in the first and fifth spaces and the block of chest wall then excised in continuity with the breast after the method of Urban; a fascial graft is used to close the defect. Or it may be done with preservation of the pectoralis muscle, when the latter is used to close the gap in the chest wall, in the way described by Handley, Patey, and Hand. The papers quoted supply further details of these, as yet, unproven operations.

Post-operative Treatment in Breast Operations

Smooth post-operative progress after breast surgery depends on perfect hæmostasis during the operation and adequate rest after it. Even after small procedures the breast heals better if both the patient and her pectoralis major are kept quiet. After radical surgery in particular, early movement of the shoulder joint not only causes pain but it prevents adhesion of the flaps to the underlying chest wall and favours the exudations of fluid. Only after the seventh post-operative day should active exercises be encouraged and passive movements with stretching have no part at any stage of management. Collections of fluid under the flaps, a nuisance much diminished by suction drainage, should be evacuated as often as they are detected either by aspiration or by sinus forceps. Marginal necrosis of flaps is seen fairly frequently by surgeons who cut their flaps thin. If the area of necrosis is of any size, it should be cut away with scissors as soon as it has demarcated. The resulting raw area may then be cleaned up with small Eusol compresses, treated with local applications of tyrothrycin and grafted. Where the original operation has required skin grafting, surplus pieces of skin may be preserved in a refrigerator at 4°C. in the container illustrated (Fig. 202) so that fresh grafts need not be cut to deal with marginal necrosis.

Minor degrees of swelling of the arm are common after radical breast operations and do not impede the full use of the arm. Frank brawny arm occasionally occurs and rarely it reaches very disabling proportions. Elevation of the arm at night on a pillow, and if possible at intervals by day, coupled with the use of an elastic arm stocking, is perhaps the most useful measure. Operative treatment by lymphangioplasty, whereby Sampson Handley sought in 1908 to form new lymph channels by passing silk threads subcutaneously on long probes from the dorsum of the hand to the chest wall, has proved

disappointing. Massive infiltration by dilute solutions of hyaluronidase, though markedly decreasing the size of a brawny arm, has a fleeting effect. Where a brawny arm is enormous and crippling, it would be worth considering treatment by excision of subcutaneous tissues and replacement of the skin on the muscles, as is done on occasion with elephantiasis of the legs. Operations such as this must however be reserved for arms which have

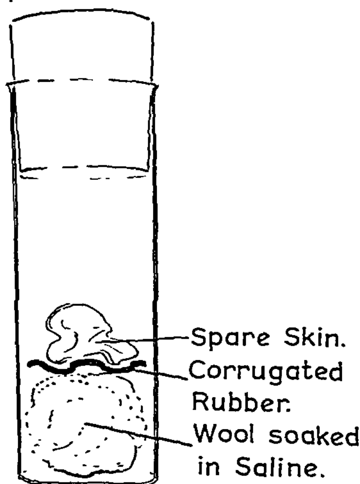


Fig. 202. Preservation of spare skin following skin grafting. The container is placed in a refrigerator at 4°C.

become virtually useless, because the resulting limb would be cosmetically hideous and would require always to be covered with clothing on social occasions. I count myself fortunate in having had no experience of such extreme measures.

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CHAPTER VIII

THORACIC WALL, PLEURA, LUNG AND BRONCHUS, MEDIASTINUM

T. Holmes Sellors

INTRODUCTION

A simple conception of the mechanics of the thorax is to regard the pleural cavities as exercising a balanced pull on the mediastinum. The mediastinum, which is much more mobile than commonly supposed, is held centrally unless the tension in one lung or pleural cavity is altered. The elastic lungs tend to retract, but are held out by negative or subatmospheric pressure of the pleural sacs against the chest wall. In the absence of pleural adhesions the degree of expansion of the lung is adjusted to the movements of the thoracic cage. For example, when a patient is breathing with the diaphragm the lower parts of the lungs will be more in use than the apex. The lung follows the chest wall excursion and this factor is made use of in physiotherapy where active inspiratory movements of the chest wall are used to restore or encourage lung expansion. The average man tends to breathe more with the diaphragm and lower ribs than women and young children; this is particularly noticeable when the ribs become rigid or when there is a tendency to emphysema or bronchospasm.

The production of a subatmospheric pressure in the pleural cavity can best be pictured when it is realized that the growing pleural sac enlarges at a much greater rate than the lung. The pleural surfaces are in apposition, like two damp plates of glass held by capillarity—able to slide against each other but firmly held. Since there is no pleural space in normal life the pressure strictly cannot be measured, but a figure of − 5 to − 10 cms. water is a fair estimate during ordinary quiet breathing. If the pleural cavity is opened air enters the space till atmospheric pressure is reached. In consequence, the lung retracts and not only is its ventilating surface lost but the balance on the mediastinum is upset. This phenomenon of open pneumothorax was largely responsible for the late development of chest surgery.

The complicated mechanisms which ensue from an open pneumothorax depend on the mobility of the mediastinum; once the balance is upset this septum is drawn towards the normal side and encroaches on the sound lung. During inspiration the outward movement of the chest wall and descent of the diaphragm increase the dimensions of the sound lung and at the same time drags even further on the mediastinum. When the thorax expands air is drawn through the trachea into the sound lung, and at the same time some air will be sucked from the collapsed side which in consequence will become smaller. The reverse processes occur during expiration; the mediastinum becomes more central and some gases are blown into the collapsed lung as well as up the trachea. There are thus three distinct mechanisms at work in any open pneumothorax:

(1) The collapse of lung.

(2) The to-and-fro movement of the mediastinum, the so-called mediastinal flap or "flutter."

(3) Paradoxical movement of the collapsed lung. With this is associated the swing of gases to and from one lung to the other, the so-called "Pendelluft." This causes a steady decrease in oxygen and a rising concentration of carbon dioxide.

Clinically, *paradoxical movement* can occur whenever the pleural cavity is opened or if a large section of chest wall becomes loose, as with multiple fractures. In many cases this paradoxical movement and open pneumothorax is tolerated and it should be recalled that a number of intrathoracic operations were at one time performed only under high spinal or local anæsthetic without too great difficulty. This "benign" or clinically

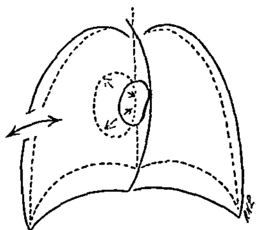


FIG 203. Right open pneumothorax. Continuous line indicates inspiration; dotted line indicates expiration Note adverse movement of mediastinum and paradoxical movement of the collapsed right lung

uneventful, paradoxical movement is not important if the patient's respiratory function is good. But if additional mechanical factors, such as obstruction of bronchi with subsequent atelectasis and severe coughing are added, a condition of highly dangerous or "malignant" paradoxical movement results and this is associated with acute respiratory distress, rising pulse rate, shock and possibly death unless the condition is controlled.

Mobility of the Mediastinum. The extent of movement of the mediastinum is a feature whose importance is not always appreciated. In a normal adult the apex of the heart may move as much as 2–3 in. when turning from one side to another; in women the displacement when moving from the erect into a lateral posture is between 400–600 ml., while in men it ranges from 550–800 ml. This is considerably larger than is commonly realized and can be made use of in the treatment of certain conditions. The term mediastinal rigidity, or fixation, gave a false impression and only occurs with gross pleural pathology; in many elderly men who undergo major thoracic operations the mobility of the mediastinum is almost as great as it is in children.

Open Pneumothorax. The open pneumothorax caused by surgical intervention is controlled during the operation by raising the pressure of gases in the trachea, thus keeping the mediastinum stable and the sound lung well ventilated. At the end of the operation active inflation of lung is carried out by the anæsthetist while the incision is being closed. Then, if a closed or water-seal drainage system is used the pleural mechanics are imitated during such time as any air or fluid in the chest drains away and does not return. The handling of a water-seal is one of the most important factors in the after-

treatment of chest operation; the tubing system must not become blocked. If there is free drainage there will be a suck back of fluid for several inches up the end of the drainage tube and this column will swing with each movement of the chest (Fig. 204).

An external opening is not the only way in which air can enter the pleural cavity; a tear in lung which may arise from injury or disease will also produce a pneumothorax and on coughing and straining air will continue to be squeezed out as long as the opening remains unsealed. A steady accumulation of air within the pleural sac after having collapsed the lung then starts to displace the mediastinum, giving rise to pressure or *tension pneumothorax*. The recognition of tension pneumothorax is not always made as readily as it should be and particularly in children displacement of mediastinum with signs of distress should always make one aware of the possibility of this occurrence. The remedy is straightforward; immediate aspiration will relieve the tension and, if a more permanent form of aspiration is required, a small needle thrust into the pleural cavity or the insertion of a small cannula and tube attached to a water-seal for the evacuation of air will allow the enclosed air to escape.

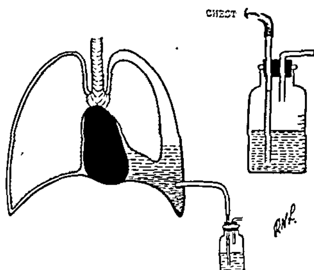


FIG. 204 Closed water-seal drainage of the pleural cavity Pleural contents are removed without any risk of air entering the cavity.

Anæsthesia. Intrathoracic surgery is closely bound up with problems of anæsthesia and control of open pneumothorax in modern anæsthetic methods enables the surgeon to operate deliberately in the presence of a widely open chest at the same time as the patient is fully oxygenated. The original "unterdrück" or under-pressure form of anæsthesia was abandoned early in the history of chest surgery, and its place was taken by "over-pressure" methods which have been modified so that the actual pressure used is not unduly high. The methods of anæsthesia used vary, but they are mostly based on endotracheal intubation after use of basal narcotics and the maintenance of anæsthesia with gas-oxygen mixtures. Controlled respiration in which spontaneous respiratory activity is temporarily abolished by the anæsthetist affords the surgeon a field of operation that is relatively immobile and facilitates the finer forms of manipulation.

Pleural Cavity. The thin, serous membrane which lines the thorax and is reflected over the lung is not attached with uniform tenacity. The visceral pleura is for practical purposes an integral part of the lung surface and cannot be removed without damage to the lung substance. The parietal pleura, though firmly adherent to the diaphragm, is very loosely attached to the chest wall and mediastinum and can be stripped off without undue difficulty except possibly at the apex. Pathological lesions increase the degree of fixity of the pleura to the chest wall, but one of the surgical assets of the pleura is that it acts as a limiting membrane or barrier to the spread of infection or tumour extension. A lung growth will spread a considerable distance over the parietal pleura before extending into the actual chest wall.

(3) Paradoxical movement of the collapsed lung. With this is associated the swing of gases to and from one lung to the other, the so-called "Pendelluft." This causes a steady decrease in oxygen and a rising concentration of carbon dioxide.

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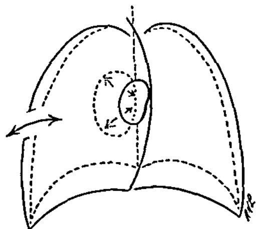


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patient must be given a head-down tilt of more than that angle. In the lateral position the trachea is almost horizontal though with the slight displacement of the mediastinum under gravity some head-down tilt will be necessary. But, if the basal bronchi of the underlying lung are to be drained this tilt must be exaggerated to an almost impossible position or the patient placed face-down with the buttocks at a higher level than the head. Drainage of lobar bronchi can only be studied in relation to the anatomy. The basal or lower lobe bronchi inevitably require inversion of the patient; while drainage of the middle lobes necessitates a supine position with the head downwards. The upper lobe bronchi drain spontaneously with the patient in the erect posture. These points have to be borne in mind in any case of chest disease where there are free secretions that cannot be cleared adequately by coughing.

SURGERY OF THE CHEST WALL AND PLEURAL CAVITY

Deformities of Chest Wall

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The only other deformity that demands surgical intervention is a large, *bony defect* in the thorax through which lung herniates. Mobilization of a rib so as to bridge the gap usually suffices if the condition is serious enough to demand attention. Alternatively metal mesh or plastic plates can be used to give sufficient support to the chest wall.

Fracture of Ribs and Sternum

There are two aspects of fracture of rib which differ widely. In the more simple form the rib or ribs are broken but the fragments are not displaced and there is minimal damage to deeper structures. In the other form the fracture is associated with physiological changes and with severe internal damage which is much more important than the broken ribs.

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The normal pleural surfaces are always moist and a balance is maintained between fluid formation and its absorption. Irritants and inflammatory processes upset this balance and cause an excess of exudate which, in obedience to the physical laws of the pleural cavity, shows a broad base and a thin film at the periphery. If air is admitted a horizontal fluid level is immediately formed. The classical shape of an enclosed pleural effusion with the patient in the erect posture can be altered by changing the patient's position so long as there are no adhesions. Once the inflammatory process has subsided absorption will occur, but the speed of resolution will depend on a variety of factors.

The majority of effusions have a high fibrinogen content and after aspiration undergo "gel" formation. The deposition of fibrin occurs more heavily on the parietal pleura than on the lung surface. This fibrin delays absorption and produces adhesions between the pleural surfaces, and later organizes into fibrous tissue with firm pleural adhesions and even complete symphysis between the two layers. Any inflammatory process or deposit of fibrin restricts the movement of the chest wall and affords one of the earliest diagnostic signs of disease. Recovery of function can only be obtained by making the chest wall move in spite of the restricting pleural pathology, and thus encourage lung movement.

Bronchial Secretions. The daily volume of bronchial secretion is greater than generally supposed. Normally bronchial movement and the action of cilia keep the air passages clear. In addition, there is the explosive blast of the cough which carries excess secretions from far down the lung to the surface in one single violent action. The ease of clearing the bronchial tree depends largely on the viscosity of the sputum; in the early stages of bronchial inflammation secretion is very viscid and only ejected with difficulty after prolonged bouts of coughing. Later the secretions are more fluid and if not excessive can easily be removed by coughing, but if the daily quantity is more than one to two ounces postural drainage may save the patient the strain of prolonged bouts of coughing.

The amount of sputum expectorated each day should be measured carefully in any thoracic case just as the urine output is recorded in cardiac or renal disease. There are, however, factors which may influence the apparent measurement. Children, for example, swallow their sputum and rarely expectorate before the age of 5 or 6 years; again, many adults swallow a large proportion of their sputum. Measurement in such cases is best made by gastric lavage first thing in the morning, when it may be found that there is a considerable quantity of sputum that has been swallowed during the night. Also, the question of secretions from the naso-pharynx being confused with that coming from the lung has to be considered. A patient with a free, purulent discharge running back from the nose may produce a considerable amount of muco-pus which can easily be added to sputum from the lungs.

Postural Drainage. Postural drainage of the bronchial tree plays a large part in all aspects of thoracic disease when the normal methods of secretion ejection are not effective. With the patient in the supine position the angle of the trachea to the horizontal is about 20 degrees. This implies that if secretions are to be removed by gravity from the trachea the supine

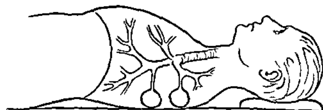


FIG 205 Common sites for formation of lung abscess In this position the pus cannot drain out. The patient would have to be placed face down.

patient must be given a head-down tilt of more than that angle. In the lateral position the trachea is almost horizontal though with the slight displacement of the mediastinum under gravity some head-down tilt will be necessary. But, if the basal bronchi of the underlying lung are to be drained this tilt must be exaggerated to an almost impossible position or the patient placed face-down with the buttocks at a higher level than the head. Drainage of lobar bronchi can only be studied in relation to the anatomy. The basal or lower lobe bronchi inevitably require inversion of the patient; while drainage of the middle lobes necessitates a supine position with the head downwards. The upper lobe bronchi drain spontaneously with the patient in the erect posture. These points have to be borne in mind in any case of chest disease where there are free secretions that cannot be cleared adequately by coughing.

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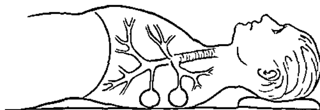


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Simple fracture of a rib resulting from direct violence, such as a kick or a blow, is accompanied by pain and bruising. Crush or "spring" injuries are liable to affect more than one rib and occur at the anterior or posterior angles, or at the maximum point of lateral convexity. The intercostal muscles help to splint the ribs and prevent displacement which only occurs in cases of extreme violence. The most serious form of injury is the *"stove-in" chest* in which multiple fractures front and back produce a loose segment of chest wall. This segment, freed from continuity with the rest of the thorax, moves paradoxically and may cause severe disturbance of the mechanics and physiology of the chest.

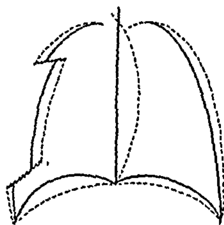


Fig. 206. "Stove-in" chest. The loose segment of chest wall moves paradoxically. Dotted line indicates inspiration, continuous line indicates expiration.

A sharp spike of rib, if depressed or pushed inwards, will puncture pleura and possibly lung and may cause pneumothorax, effusion or hæmothorax. The practice of applying firm strapping to the chest wall over a rib fracture can easily produce these complications and should be abandoned. The treatment of a simple fracture is to infiltrate the area with local anæsthetic and apply heat or similar measures for the relief of pain, and to encourage active movements of the chest at the earliest moment.

Internal complications of broken ribs require more urgent treatment than the fracture itself. Hæmorrhage from torn or punctured intercostal vessels may produce an extensive hæmothorax and if the lung surface is injured escape of air can lead to tension pneumothorax.

A separate consideration is the management of the "stove-in" chest. Here contusion and damage to lung is probable and retained secretions and atelectasis contribute to dangerous paradoxical movement. Immediate treatment consists of trying to immobilize the loose segment of chest wall with strapping applied over a firm pad or moulded slab of plaster of Paris. This, however, can only depress the fragment and produce permanent deformity. Consequently, as soon as circumstances permit, immobilization by open operation and suture or by traction should be considered. Open operation is reserved for cases in which the chest has to be opened for other injuries or for compound fractures. It is not always easy to suture the loose section of chest wall firmly in its original position. The alternative method is effected by placing two or three towel clips round the ribs in the loose segment and exerting weight traction for 10–14 days; by this time some degree of union or fixity will have occurred. During this period air and blood should be completely removed from the pleural cavity and retained bronchial secretions which predispose to atelectasis aspirated bronchoscopically.

Diseases of Ribs

Osteomyelitis of rib can occur as the result of metastatic infection and carries with it the risk of infecting the pleural cavity. Secondary infection of rib is uncommon, but can result from pleural or pulmonary infection. It is, however, surprisingly rare to find infection of the cut ends of rib in the case of empyema drainage after rib resection. Typhoid osteomyelitis of rib and sternum is a fairly common late complication of the

specific enteritis and if the nature of the infection is not recognized further outbreaks of typhoid fever may follow.

Tuberculosis of rib is a condition whose pathology is sometimes obscure. Metastatic involvement can occur but the so-called "primary" tuberculosis of rib is usually a secondary phenomenon consequent on a cold abscess from a breaking down intercostal or internal mammary gland. This spreads along the intercostal groove and secondarily produces a periostitis. The subcutaneous cold abscess that presents in these cases is at the site where the perforating branches leave the intercostal channel. Excision of eroded or necrotic rib may fail in its object as the primary focus in the glands (often some distance away) remains untreated.

Benign tumours of rib in the form of chondroma or osteoma are expanding, fusiform masses with a tendency to project internally rather than towards the surface. Local excision will suffice in most instances but some tumours, frequently associated with low grade fever, tend to recur and show malignant characteristics.

Among primary malignant tumours of the bony thorax sarcoma and Ewing's tumour are the most common. Neither wide excision, nor radiotherapy, achieve lasting success save in exceptional cases. Secondary invasion of rib by lung, prostate or breast growths are rarely suitable for treatment though some palliation of pain can be obtained by radiotherapy or endocrine therapy. Local excision is sometimes indicated.

Pneumothorax

The presence of air in the pleural cavity leads to passive collapse of the underlying lung to a degree that depends on the pressure and amount of the air in the pleural cavity. The two methods by which air can enter are through a communication with the outside air through the chest wall, or from rupture of the lung. *Artificial pneumothorax* is the best example of the deliberate admission of air through a needle and the degree of collapse is controlled by the amount of air that is introduced at suitable intervals. The rate of absorption of the enclosed gases varies from case to case and depends largely on the permeability of the pleural membrane and any inflammatory reaction that may have occurred. A penetrating wound of the chest wall which does not break the surface of the lung will admit air in increasingly large volumes if the surface wound remains open. Air is sucked in with each breath and the pressure of the enclosed air not only collapses lung but brings about displacement of the mediastinum—pressure or tension pneumothorax. The treatment of "*sucking*" *wounds* is their immediate closure either by pad (in the form of first-aid dressing) or closure of the wound following deliberate surgical excision. The enclosed air must be removed.

Spontaneous pneumothorax which arises from rupture of lung tissue is often produced by ulceration of tuberculous lesions, and infection of the pleural space by tubercle bacilli is likely to occur. Formerly this was said to be the most common form, but in recent years spontaneous pneumothorax has been recognized as being caused by rupture of lung surface occasioned by bursting of a small bulla or an emphysematous cyst. The origin of small bullæ is obscure; many occur on the fringes of the lung and result from obstruction of a peripheral bronchus or bronchiole, the bulla being to all intents and purposes a small, emphysematous cyst. This is seen sometimes as a consequence of tuberculous scarring when occlusion of a bronchus is not complete but is sufficient to cause distension rather than collapse.

The onset of spontaneous pneumothorax is accompanied by pain and shortness of breath. This may be severe but is often insidious and occurs in a healthy person following some movement such as throwing a stone or suddenly stretching overhead. On examination, increased resonance of the percussion note, absence of breath sounds and vocal fremitus suggest the presence of air in the pleural cavity. This will be confirmed by radiography. Many of these cases resolve spontaneously, the air being slowly absorbed,



(*a*) (*b*)

FIG 207. Chronic pneumothorax.

(*a*) Large pneumothorax space at apex of lung, still adherent, and collapse of the lower lobe.
(*b*) Following suction drainage showing partial expansion of lower lobe and the appearance of cystic spaces

but recurrence is common and further measures have to be adopted if repeated lung collapse leads to incapacity. Most pneumothoraces in young people do not progress to tension phenomena, but in the more elderly, where there is degenerative emphysema or cyst formation, the opening in the lung surface does not close and the pneumothorax persists. In these chronic cases aspiration of air should be carried out and the pleural pressures watched to see if there is a continuous leak between the lung and pleural cavity.

When a pneumothorax is recurrent or *chronic*, treatment resolves itself into trying to obliterate the pleural space by inducing a chemical pleurisy or to suturing the open breach in the lung surface. Chemical pleurodesis is performed by introducing 1 per cent iodine mixed in talc powder through a thoracoscope. The powder is blown in through an insufflator on to the surface of the lung, after which all air is quickly removed by continuous suction. The irritation of the pleura causes adhesions to form and prevents the lung from collapsing. The procedure is painful and the patient requires considerable sedation over the first few days. Injection of silver nitrate is also recommended. It is successful in some cases.

In chronic cases, thoracotomy may be the only means by which success will be achieved. The chest is opened and the opening identified; this is not usually suitable for suture and

the cystic area if possible should be excised locally. This, however, will only be practicable if the damaged area is localized; a degenerative emphysematous lung or lobe may have to be excised if cure is to be obtained and in this type of case the difficulties of age and the state of the lungs have to be taken into account. Indeed, in some cases little can be done except to leave the pneumothorax in its chronic state. Prior to major surgical intervention, full examination of the lungs by bronchoscopy, bronchography, and bronchospirometry should be undertaken before operation is contemplated.

Hæmothorax and Penetrating Wounds

The presence of blood in the pleural cavity is a common occurrence in warfare resulting from penetrating wounds when the vessels in the chest wall or lung are damaged. When the blood is removed it is found to be liquid in the majority of cases. This phenomenon caused a great deal of confusion until it was realized that the blood clots quite normally, but the movement of the heart and lung agitates the clot to such an extent that defibrination occurs. In cases of penetrating injuries the defibrination is complete within an hour or so. The proof of this lies in the fact that liquid blood aspirated from the pleural cavity contains no fibrinogen. Another feature is that the hæmoglobin concentration is 30–40 per cent below that of the circulating blood. In other words, blood has been diluted by an exudate which appears as the result of pleural irritation. If the blood is not removed, the exudate continues to form and in the course of a week or 10 days produces enough fibrin to allow the residual blood and effusion to clot.

This so-called *clotted hæmothorax* is a secondary phenomenon and is strictly not due to clotting of the blood, but to clotting of the fibrin which is heavily blood-stained. The hæmoglobin percentage of a hæmothorax after the first 2 or 3 days rapidly falls to 10 per cent or 15 per cent as a result of dilution and the error of regarding a three or four pint "hæmothorax" as indicating a loss of three or four pints of blood from the circulation must be avoided.

Pathologically, a hæmothorax may develop as a result of tuberculosis of the pleura or from new growth; nearly all other cases are traumatic in origin and many cases of pleural effusion which are found to be slightly blood-stained have their origin in damage to a blood vessel by the exploring needle.

In the case of penetrating wounds, a hæmothorax is usually complicated by the presence of air which gains access through the chest wall or from damaged lung. A patient who suffers from such an injury will invariably be distressed and breathless according to the amount of blood and air in the pleural cavity, and will furthermore have bloody sputum as a result of the lung injury. Pain and weakness inhibit coughing and atelectasis is a common complication. The treatment of such cases is to remove enough air and blood as an emergency to allow the mediastinum to become central and reduce the grosser intrathoracic pressure effects, and to endeavour to keep the bronchi free from secretions by coughing or, failing that, by bronchoscopy. It is an essential part of the first-aid treatment that any open wound on the surface should be closed and rendered airtight, either by temporary dressings or by more definitive surgery. With all penetrating wounds the risk of later infection is present, though the use of antibiotics has overcome a great deal of this problem.

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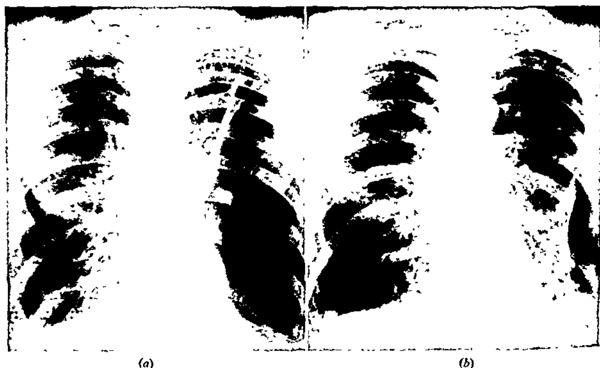
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In chronic cases, thoracotomy may be the only means by which success will be achieved. The chest is opened and the opening identified; this is not usually suitable for suture and

the cystic area if possible should be excised locally. This, however, will only be practicable if the damaged area is localized; a degenerative emphysematous lung or lobe may have to be excised if cure is to be obtained and in this type of case the difficulties of age and the state of the lungs have to be taken into account. Indeed, in some cases little can be done except to leave the pneumothorax in its chronic state. Prior to major surgical intervention, full examination of the lungs by bronchoscopy, bronchography, and bronchospirometry should be undertaken before operation is contemplated.

Hæmothorax and Penetrating Wounds

The presence of blood in the pleural cavity is a common occurrence in warfare resulting from penetrating wounds when the vessels in the chest wall or lung are damaged. When the blood is removed it is found to be liquid in the majority of cases. This phenomenon caused a great deal of confusion until it was realized that the blood clots quite normally, but the movement of the heart and lung agitates the clot to such an extent that defibrination occurs. In cases of penetrating injuries the defibrination is complete within an hour or so. The proof of this lies in the fact that liquid blood aspirated from the pleural cavity contains no fibrinogen. Another feature is that the hæmoglobin concentration is 30–40 per cent below that of the circulating blood. In other words, blood has been diluted by an exudate which appears as the result of pleural irritation. If the blood is not removed, the exudate continues to form and in the course of a week or 10 days produces enough fibrin to allow the residual blood and effusion to clot.

This so-called *clotted hæmothorax* is a secondary phenomenon and is strictly not due to clotting of the blood, but to clotting of the fibrin which is heavily blood-stained. The hæmoglobin percentage of a hæmothorax after the first 2 or 3 days rapidly falls to 10 per cent or 15 per cent as a result of dilution and the error of regarding a three or four pint "hæmothorax" as indicating a loss of three or four pints of blood from the circulation must be avoided.

Pathologically, a hæmothorax may develop as a result of tuberculosis of the pleura or from new growth; nearly all other cases are traumatic in origin and many cases of pleural effusion which are found to be slightly blood-stained have their origin in damage to a blood vessel by the exploring needle.

In the case of penetrating wounds, a hæmothorax is usually complicated by the presence of air which gains access through the chest wall or from damaged lung. A patient who suffers from such an injury will invariably be distressed and breathless according to the amount of blood and air in the pleural cavity, and will furthermore have bloody sputum as a result of the lung injury. Pain and weakness inhibit coughing and atelectasis is a common complication. The treatment of such cases is to remove enough air and blood as an emergency to allow the mediastinum to become central and reduce the grosser intrathoracic pressure effects, and to endeavour to keep the bronchi free from secretions by coughing or, failing that, by bronchoscopy. It is an essential part of the first-aid treatment that any open wound on the surface should be closed and rendered airtight, either by temporary dressings or by more definitive surgery. With all penetrating wounds the risk of later infection is present, though the use of antibiotics has overcome a great deal of this problem.

The *treatment* of the actual hæmothorax consists in early and complete removal of all

the liquid and air in the chest. One of the chief lessons learnt during the last war was that if these cases were completely aspirated within 24–48 hours of the injury the complications were negligible and the restoration of function rapid. Aspiration has to be performed without admitting any air and it is an operation that may well take half to three-quarters of an hour. If it is not completed at the first attempt it should be repeated on the following day or days until no further air or fluid can be removed (vide Aspiration, p. 414). In cases in which treatment is delayed for any reason the problem of secondary clotting and infection exists. Pleural decortication plays an important part in dealing with clotted fibrinothorax; the blanket-like layers of fibrin are removed from the lung and chest wall and early re-expansion of lung and lung function is obtained. If infection has occurred the steps taken follow those used in the handling of empyema, namely antibiotics for sterilization and, if that fails, drainage, accompanied by vigorous breathing exercises to encourage lung expansion.

It was a general policy during the last war to remove all intrathoracic foreign bodies whose diameter was more than 1·5 cm. This can be effected either during thoracotomy, if this is carried out immediately after injury, or at a later stage through a small incision directly over the missile. All foreign bodies lie very close to the lung surface and are never deeply buried in lung substance. This position is a result of rebound from the far side of the chest wall, the mediastinum acting as a rigid structure in most cases. The missile, having traversed the thorax, is held up by the ribs on the far side if it does not have sufficient velocity to make its exit.

Associated lung lesions in cases of penetrating or crush injuries of the chest wall and pleura take a variety of forms. Puncture or "through and through" wounds cause some bleeding from the lungs with extravasation into the surrounding tissues which helps to arrest hæmorrhage. More massive laceration of lung may even lead to partial amputation, but the usual crush picture is that of a large, diffuse hæmatoma which takes 4–6 weeks to resolve. Mucus and blood in the bronchial tree predispose to atelectasis which complicates the picture.

High explosion or "blast" injuries produce a definite pattern of thoracic injury. The periphery of the lungs and particularly the lower fringes are affected by patchy effusions and hæmorrhages which resolve comparatively quickly (within a week or 10 days) if there are no other complicating factors. After immediate resuscitation measures, which include oxygen therapy and bronchoscopy to remove bloody mucus, little active treatment is required though should any anæsthetic be required inhalation methods must be avoided owing to the diminished ventilating surface of the lung.

In all cases of extensive injury the risk of damage to extrathoracic structures is present. Abdominal trauma can easily be overlooked in the early stages and rupture of the diaphragm with tearing or penetration of liver, spleen, stomach, and colon is encountered in a proportion of cases. When abdominal injury occurs its treatment takes priority over thoracic lesions though with the use of combined thoraco-abdominal incisions both serous cavities can be explored at the same operation.

Pleural Infection

Inflammatory changes in the pleura follow normal lines but with the added complication that the area potentially infected is the whole of the pleural cavity. The ordinary processes of irritative exudate, localization, and abscess formation are still encountered,

but the mechanics of the pleural cavity produces additional problems which affect treatment.

Pleuritis usually follows infection of the underlying lung and the early signs and symptoms tend to be overshadowed by the primary cause in the lung. It has been found convenient to divide the stages of infection into two phases, that of diffuse suppurative pleurisy and that of the localized pleural abscess or empyema.

Diffuse Suppurative Pleurisy. The onset is characterized by an active exudate with varying fibrinogen content and of varying volume. For example, streptococcal infections are usually large in volume and with a low fibrin content, while the pneumococcal pleuritis produces masses of fibrin and is not usually of large size. After the first 2 or 3 days fibrin becomes deposited from the fluid and the cellular elements of pus become visible. The fluid which originally was clear and contained actively growing organisms becomes turbid and then frankly purulent during the course of 4–7 days. Fibrin is deposited on the pleural surfaces, particularly the parietal pleura and at the edges of the effusion, and when this process is complete the infection which originally could have involved the whole cavity becomes localized.

Localized Pleural Abscess. A localized abscess is now developing and strictly to this phase the term empyema is reserved. As time progresses the fibrin organizes, becomes more firm and the collection of pus more clearly circumscribed. Later, if not treated, the condition passes into the chronic stage in which there are rigid, fibrous walls to the abscess. These fibrous walls restrict the movement of the chest and bind down the lung preventing its expansion. Still later, cicatrization will lead to an immobile and permanently contracted area of chest wall and restricted lung function. If this process occurs over the whole of one side the term "frozen chest" is sometimes applied.

Formerly considerable distinction was made between the types of inflammation and the terms syn- and meta-pneumonic were used to describe an empyema that occurred synchronously with the lung inflammation in the first instance and that which occurred at an interval after the pneumonia in the latter. Streptococcal infections were the best examples of the first group, also some pneumococcal conditions in children, but the characteristic empyema that followed the pneumococcal lobar pneumonia was the classical example of the meta-pneumonic form. During the past few years the whole picture of pleuro-pneumonic inflammation has changed, largely owing to the influence of antibiotics. Lobar pneumonia is practically never seen and if properly treated is almost a passing indisposition. Similarly, other forms of lung inflammation have become transitory mild affections. The former florid type of empyema has given place to a much more insidious form in which the pleural contents are frequently sterile or at any rate of only low infectivity.

The signs and symptoms of empyema are well recognized, but it is surprising how frequently the condition remains undiagnosed until a late stage, the most important factor being the masking of the condition by the primary lung inflammation. Physical signs are often equivocal, particularly in children, and accurate diagnosis *usually depends* on radiological findings and the result of pleural aspiration. The *only certain* method of diagnosis is the recovery of purulent fluid from the pleural cavity.

Types of Empyema. The common form of empyema is posteriorly placed at the base and spreading forwards towards the axilla, gravity and the normal erect or recumbent position of the patient determining the site. If the fluid is present in large quantities or

the liquid and air in the chest. One of the chief lessons learnt during the last war was that if these cases were completely aspirated within 24–48 hours of the injury the complications were negligible and the restoration of function rapid. Aspiration has to be performed without admitting any air and it is an operation that may well take half to three-quarters of an hour. If it is not completed at the first attempt it should be repeated on the following day or days until no further air or fluid can be removed (vide Aspiration, p. 414). In cases in which treatment is delayed for any reason the problem of secondary clotting and infection exists. Pleural decortication plays an important part in dealing with clotted fibrinothorax; the blanket-like layers of fibrin are removed from the lung and chest wall and early re-expansion of lung and lung function is obtained. If infection has occurred the steps taken follow those used in the handling of empyema, namely antibiotics for sterilization and, if that fails, drainage, accompanied by vigorous breathing exercises to encourage lung expansion.

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essence of obliterating the pleural space. A two-way tap attached between the needle and syringe overcomes this difficulty and the apparatus should be as carefully chosen as any complicated surgical apparatus before starting the operation. Both patient and operator should be comfortable because the aspiration of a pint of pus or more may take 15–20 minutes. Local anæsthetic is used in the skin and in the chest wall superficial to the pleura. Through this area, which will have been carefully located by clinical and possibly radiological examination, the needle is passed. Once it has entered the pleural



FIG. 209. Pleural effusion.
(*a*) Large left-sided effusion with slight displacement of mediastinum.
(*b*) Following aspiration

cavity (verified by the withdrawal of pus) a light clamp is placed on the needle flush with the skin to prevent it being thrust too deeply and injuring underlying lung. Too rapid removal of pleural fluid may cause some distress and coughing which will subside if a short period of rest is allowed. If the fluid is at all thick or if there are flakes of fibrin the needle may become blocked and care has to be exercised in freeing the obstruction. If the blocking occurs repeatedly when a moderately wide bore needle is being used this is an indication that the pleural space has become thick-walled and the empyema localized. The thickness of the pleural membrane can be easily judged by an experienced operator. If he feels that he is thrusting the needle through wet cardboard or blotting paper the chances are that the pleural infection will have started to become localized. Aspiration should always be as complete as possible and persisted with until nothing more can be withdrawn. It should be repeated as soon as any fluid or pus re-collects, possibly the next day. Several days should not be allowed to elapse if any accumulation of fluid is recognized.

While fibrin is being deposited on the walls of the cavity there are fibrin flakes floating loose in the effusion. In the early stages the deposit is soft and friable, but at the end of 10 or 14 days it attains the thickness of several millimetres. In other words, localization

if for any reason air is admitted into the pleural cavity a total empyema results. Posterior pockets which take a rounded shape are common, as also interlobar effusions which may lie anywhere along the line of one of the fissures. The most confusing and probably the most common site of an interlobar effusion is anterior, presenting on the surface under the level of the nipple. Mediastinal collections are often quoted but these are extremely rare. So-called mediastinal empyemata are often misnomers for some other condition, such as atelectasis of a lower lobe.

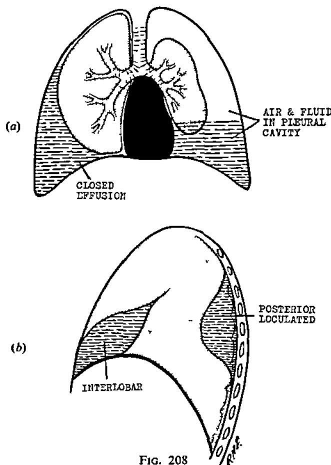


FIG. 208

(a) Right side shows the outline of a "closed" pleural effusion On the left the presence of air leads to a horizontal fluid level

(b) Lateral view showing the anteriorly placed pyriform appearance of an interlobar effusion and a posterior loculated pocket.

Treatment. The principles underlying treatment are the same as in any other case of inflammation. First, the sterilization or removal of the products of inflammation and secondly, the closure of the abscess cavity. It is the second feature to which too little attention is given. In most parts of the body the soft walls fall together after the abscess has been drained, while in the chest there is a rigid outer wall and an inner one which do not readily re-expand to fill up the space. The methods of treatment depend also on the stage of the inflammation, the treatment for the diffuse stage being quite different from that for a localized abscess.

Antibiotics. In many cases the patient will have been treated by antibiotics before the pleural effusion has developed or been recognized. This accounts for the increasingly large number of sterile, yet purulent effusions that are encountered at the present time. Once an effusion is recognized, careful diagnostic aspiration is undertaken and, from the moment that this is performed until the cavity in the pleura is completely obliterated, there should be no break in continuity of treatment. Errors creep in through inefficient treatment in the early stages and in the hiatus that occurs between the so-called medical and surgical aspects of treatment. At the initial aspiration the character of the pus and its bacteriology is determined. Once this is done a formal and complete aspiration should be undertaken with the aim of removing all fluid possible and at the same time injecting penicillin or some suitable antibiotic at the completion of the process.

Aspiration. The essential feature in pleural aspiration is to avoid air entering the free pleural cavity. If this occurs air rises to the top and the apex of the lung will fall away, thus creating the worst possible conditions for re-expansion of the lung which is the

Aspiration is attempted in the early stages, but should be abandoned as soon as the needle fails to remove the contents of the pleural cavity. If fluid is accumulating rapidly an intercostal tube can be inserted through a trocar and cannula, and the tube connected to a closed water-seal system. This has the advantage of imitating the sub-atmospheric pressure of the pleural cavity and encouraging lung to re-expand at the same time as the contents are being drained. The great disadvantage of intercostal drainage is its inadequacy; fibrin masses block the tube and drainage is ineffective. Once the presence of fibrin has been established (about 2 weeks from the onset of the infection) it can be

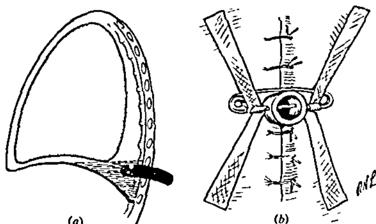


FIG 211

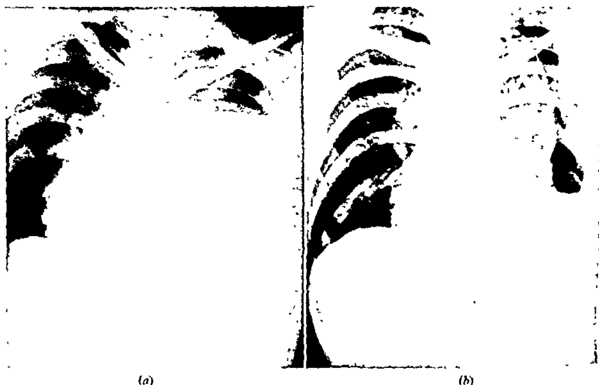
(a) Correct site for draining a total empyema The tube is placed as far back and as low as possible

(b) Method of holding drainage tube in position. The tube should actually be of much wider bore than that depicted.

assumed that the empyema can be drained without the lung tearing away from the chest wall. The empyema cavity is exposed through a small thoracotomy incision in which a short length of rib is resected. This is usually in the paravertebral line, about the ninth rib. The parietal pleura is excised and sent for histological examination, and the contents of the cavity are carefully removed and all corners of the abscess cleaned out by suction and lavage. *A large tube is then inserted at the lowest point of the cavity and carefully* placed so that it will not impinge on the underlying lung. The end of the tube, which projects, is transfixed by a safety pin placed flush with the skin and held in position by fine strips of adhesive (Fig. 211). The tube should be cut short close to the safety pin and the wound is covered by a corset dressing which should not have to be changed more than once or twice a day after the first 24 hours.

The handling of the drainage tube is the most important feature in obtaining closure of the cavity. Encouraged by inspiratory breathing exercises and the patient's own activities, the lung will slowly re-expand and the only satisfactory way in which the healing of the empyema may be judged is by the use of *pleurograms*. These consist of instilling radio-opaque oil through the tube prior to taking antero-posterior and lateral radiograms. Pleurograms should be done every 10 or 14 days once signs of re-expansion are obvious, but the tube itself should not be removed unless the shape of the cavity or irregular healing shows drainage to be inadequate. For example, sudden expansion at the base leaving a pocket higher up will require insertion of a longer tube into the upper pocket and that tube will be retained in place until the pocket is closed. Then it will be withdrawn at the rate of about 1 in. a week until healing is complete. The drainage tube

is occurring. If violent physical stresses are placed on this membrane, the lung may tear away from the chest wall and spread the area of infection. By the time there are sufficient fibrin flakes in the fluid to make aspiration difficult the margins of the cavity where the lung approximates to the chest wall are sufficiently well organized to constitute a formal abscess cavity. As has already been said, the fibrin which in the first instance is loosely deposited on the walls becomes organized and ultimately converted into fibrous tissue.



(a) (b)

FIG 210. Total empyema under tension

(a) The mediastinum is displaced to the right and there is a large air space above the fluid level. The lung is completely collapsed

(b) Appearance following repeated aspiration 3 days later Mediastinum is now central and the lung is showing some signs of re-expansion.

This in fulness of time cicatrizes and leads to contracture of the chest wall as well as being an impediment to lung re-expansion. Certain organisms produce fibrin readily, notably pneumococci and staphylococci. Streptococcal infections contain fibrinolytic enzymes which delay the process of localization, a feature that is made use of in some forms of therapy.

The clinical course of a localized pleural abscess depends on two factors; first, the control of the infection, and secondly, the efficiency of treatment. At the present time, the use of antibiotics leads to early sterilization of the empyema contents. This does not mean that the condition is cured since an infected abscess is replaced by a sterile one which is equally effective in preventing lung re-expansion. Indeed, the sterilization of an empyema removes fibrinolytic enzymes and causes a heavier deposit of fibrin than normal. Apart from this, there is the toxic factor which, if chronic, is deleterious to the patient's general condition, and many cases of apparently sterile empyema show all the features of chronic toxæmia with severe anæmia and wasting.

Drainage. As soon as an empyema has been diagnosed early treatment is imperative.

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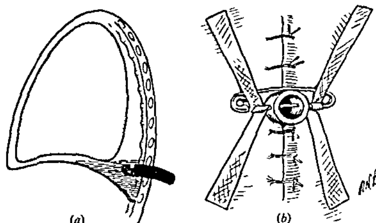


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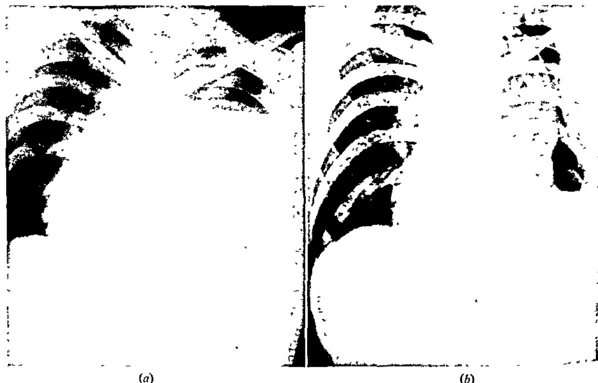


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drained by one or more tubes to which suction is applied. Drainage and suction are persisted with until the pleural cavity is finally obliterated. Decortication operations are less successful in very chronic cases but they are of value in reducing the period of hospital treatment in the early chronic cases.

The early history of empyema treatment was highly discreditable to surgery because drainage was either performed too late or carried out inadequately. Sellors and Cruickshank (1951), in a series of 622 chronic cases, found that late or inadequate drainage accounted for 62 per cent. Though chronic empyema is not often seen at the present time, the crippling deformity and ill-health that can result makes it imperative that all examples of pleural infection should be treated urgently and not be neglected. Maltreatment in the early stages may require months of energetic treatment before the condition is finally controlled.

SURGERY OF THE LUNG

Lung Abscess

Lung abscess commonly results from obstruction of a bronchus by infective material. The process is essentially that of segmental necrosis which follows the blocking of the bronchi by some infective material. Infection is the dominant factor and the segmental area, which is initially atelectatic as a result of the obstruction, proceeds rapidly to local gangrene with liquefaction and distension of the space by gas-producing anærobic organisms. In the surrounding lung tissue there is a considerable degree of pneumonitis with slow development of fibrous tissue in the wall of the abscess.

Since the abscess cannot drain as a result of the obstruction, signs of severe toxæmia develop and little or no sputum is produced. As the abscess expands and increases in size it may suddenly ulcerate into an adjacent bronchus; this is heralded by hæmoptysis and followed by a sudden discharge of the foul contents of the abscess into the bronchial tree. In other words, a patient, though toxic and without sputum, may dramatically cough up several ounces or even a pint of stinking pus in a few minutes. This form of drainage reduces the toxæmia but increases the risk of aspiration infection of lung. It cannot be regarded as an effective form of spontaneous cure. The abscess rarely ruptures into the pleural cavity because its peripheral surface lies against the visceral pleura and quickly becomes adherent to the parietes opposite it.

The *distribution of lung* abscesses depend largely on gravity. With a patient lying on the back or in the semi-sitting position the two dependent segmental bronchi are the postero-lateral branch of the upper lobe and the branch to the apex of the lower lobe (Fig. 205). Obstructive abscesses in the basal bronchi are rare because the cough reflex and expulsive movement of the diaphragm is usually sufficient to eject infective material before it becomes lodged or impacted. The role of gravity is further stressed by the fact that if men who habitually work in a crouching position or who are swimming (as with shipwrecked sailors) [illegible]

[illegible]

The clinical picture prior to the introduction of antibiotics was formidable. The source of the abscess is often suggested by foul teeth or gross oral infection, or following an operation on the throat or nose. Inhaled foreign bodies such as peanuts, toys, pins, and so on have long been recognized as possible causes of abscess, but early and efficient bronchoscopy has done much to reduce the incidence. Severe toxæmia quickly follows

is never removed from a cavity; it is only removed from a track in the chest wall. In other words, if healing is to be complete, the lung and the chest wall must approximate completely before the tube is removed.

Fibrinolytic enzymes, such as streptokinase or trypsin, can be used with effect in hastening solution of the fibrin walls, but the basis of all empyema closure is adequate drainage, plus intensive inspiratory breathing exercises. These exercises, which have to be performed for hours rather than minutes each day, are the most valuable means by which lung function is restored. As the chest wall regains its movement so does the lung show signs of re-expansion.

Chronic Empyema

An empyema cavity which persists for more than 10 or 12 weeks may be regarded as chronic and the first step is to decide whether or not the drainage is adequate.

Should the empyema persist after proper drainage, a cause other than infection should be sought; unsuspected tuberculosis and new growth are two common causes and occasionally retained foreign bodies, such as drainage tubes, swabs or bone sequestra are found. Re-exploration is almost certainly indicated if the progress of healing remains static.

When adequate drainage has been established and toxæmia reduced, the patient should be allowed out of bed and in many thoracic units the patients are encouraged to take active and intensive exercise (walking up to 10 to 15 miles a day) in the later stages of their convalescence. The blood count should be watched as secondary anæmia can develop insidiously during the course of healing. Any bleeding from the tube track may herald pressure erosion of an intercostal artery or lung. No tube should be painful; if so, it must be examined and adjusted, but the cardinal rule is not to change the tube more often than absolutely necessary. In former times, the teaching that the tube should be removed, boiled, and replaced daily was one of the most fertile sources of chronic empyema. An ordinary empyema drained at about the end of a fortnight should be healed within 6 or 8 weeks, but a chronic or maltreated case may take many months before final healing is achieved. The tendency to indulge in thoracoplastic operations should be resisted as long as pleurograms show that the cavity is steadily getting smaller, and at the present time there is practically no place for the operations that bear the names of Schede, Estlander, and Roberts. A chronic, small cavity may sometimes be treated by laying it freely open and allowing to granulate in, an operation sometimes aptly referred to as "saucerization"!

The presence of a broncho-pleural fistula complicates healing, but the majority of small fistulæ heal under ordinary drainage measures. A large fistula may require exposure and attempts at suturing, but if there is any underlying disease in the lung such as bronchiectasis, excision of lung may ultimately be required.

As an alternative to prolonged drainage, the operation of *pleurectomy* has certain advantages but it must be regarded as a major procedure. The operation consists of a long thoracotomy incision and then freeing of the fibrous, pleural abscess from the deep surface of the chest wall. Dissection is then carried round the margins of the abscess and decortication of the fibrous deposit off the surface of the lung completes the removal of the abscess. The best results from this operation are obtained in cases of large empyema which shows few signs of closing after 8–12 weeks. Once the decortication is complete, the anæsthetist endeavours to inflate the lung to its normal volume and the space is

drained by one or more tubes to which suction is applied. Drainage and suction are persisted with until the pleural cavity is finally obliterated. Decortication operations are less successful in very chronic cases but they are of value in reducing the period of hospital treatment in the early chronic cases.

The early history of empyema treatment was highly discreditable to surgery because drainage was either performed too late or carried out inadequately. Sellors and Cruickshank (1951), in a series of 622 chronic cases, found that late or inadequate drainage accounted for 62 per cent. Though chronic empyema is not often seen at the present time, the crippling deformity and ill-health that can result makes it imperative that all examples of pleural infection should be treated urgently and not be neglected. Maltreatment in the early stages may require months of energetic treatment before the condition is finally controlled.

SURGERY OF THE LUNG

Lung Abscess

Lung abscess commonly results from obstruction of a bronchus by infective material. The process is essentially that of segmental necrosis which follows the blocking of the bronchi by some infective material. Infection is the dominant factor and the segmental area, which is initially atelectatic as a result of the obstruction, proceeds rapidly to local gangrene with liquefaction and distension of the space by gas-producing anærobic organisms. In the surrounding lung tissue there is a considerable degree of pneumonitis with slow development of fibrous tissue in the wall of the abscess.

Since the abscess cannot drain as a result of the obstruction, signs of severe toxæmia develop and little or no sputum is produced. As the abscess expands and increases in size it may suddenly ulcerate into an adjacent bronchus; this is heralded by hæmoptysis and followed by a sudden discharge of the foul contents of the abscess into the bronchial tree. In other words, a patient, though toxic and without sputum, may dramatically cough up several ounces or even a pint of stinking pus in a few minutes. This form of drainage reduces the toxæmia but increases the risk of aspiration infection of lung. It cannot be regarded as an effective form of spontaneous cure. The abscess rarely ruptures into the pleural cavity because its peripheral surface lies against the visceral pleura and quickly becomes adherent to the parietes opposite it.

The distribution of lung abscesses depend largely on gravity. With a patient lying on the back or in the semi-sitting position the two dependent segmental bronchi are the postero-lateral branch of the upper lobe and the branch to the apex of the lower lobe (Fig 205). Obstructive abscesses in the basal bronchi are rare because the cough reflex and expulsive movement of the diaphragm is usually sufficient to eject infective material before it becomes lodged or impacted. The role of gravity is further stressed by the fact that if men who habitually work in a crouching position or who are swimming (as with shipwrecked sailors) dev[illegible]

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is never removed from a cavity; it is only removed from a track in the chest wall. In other words, if healing is to be complete, the lung and the chest wall must approximate completely before the tube is removed.

Fibrinolytic enzymes, such as streptokinase or trypsin, can be used with effect in hastening solution of the fibrin walls, but the basis of all empyema closure is adequate drainage, plus intensive inspiratory breathing exercises. These exercises, which have to be performed for hours rather than minutes each day, are the most valuable means by which lung function is restored. As the chest wall regains its movement so does the lung show signs of re-expansion.

Chronic Empyema

An empyema cavity which persists for more than 10 or 12 weeks may be regarded as chronic and the first step is to decide whether or not the drainage is adequate.

Should the empyema persist after proper drainage, a cause other than infection should be sought; unsuspected tuberculosis and new growth are two common causes and occasionally retained foreign bodies, such as drainage tubes, swabs or bone sequestra are found. Re-exploration is almost certainly indicated if the progress of healing remains static.

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an infective obstruction, and the patient develops a high temperature and rapid pulse rate and quickly loses weight and becomes anæmic. An irritative cough is often present, but little or no expectoration occurs in the early stages, though a foul odour may be recognized from a distance in the breath of the patient. When an abscess ruptures a flood of fœtid, stinking material, often mixed with blood, occurs and in some cases unless the patient is promptly postured he can drown in his own pus. Cerebral abscess formation and metastatic infections were common in this putrid form of lung abscess, but since the introduction of antibiotics the whole course of lung suppuration has become more benign and surgical treatment has been revolutionized.

In any lung abscess two important features have to be considered; the location of the abscess and the cause. As has been suggested, bronchial "embolism" determines the segmental distribution of the lesion. Inflammatory changes spread to adjacent parts of the lung but the central necrosis will be located in a segment. The second factor is the nature of the obstruction and the bacteriology. A few abscesses are produced by foreign bodies but these must be infected if they are to produce the associated local gangrene that is the main feature of an abscess. Specific organisms, notably the staphylococcus, tend to produce a large distension type of abscess which may resemble a cyst and which may resolve spontaneously.

In addition to infection, neoplastic changes within the lung can lead to bronchus obstruction and abscess formation, and one of the forms of lung abscess that is associated with new growth is an obstructive form developing peripheral to the tumour. This type of abscess contrasts with breaking down of growth itself in that the true abscess shows a relatively thin wall of uniform thickness, whereas the breaking-down growth shows irregular thickness of walls and an outline that is not always circular.

Once an abscess has spontaneously discharged by erosion into the bronchial tree, it usually acquires two or three small drainage orifices. If these are adequate and the contents are quickly discharged, the distension character of the cavity is removed and the abscess shrinks in size. It may then heal by the ordinary process of granulation tissue obliteration, but mechanical factors, such as a lung slough, may hinder the discharge. Autolysis leads to liquefaction of tissue and it is unusual for most of the contents not to be evacuated though persistent infection will continue to produce pus. After a few weeks, if there are several drainage orifices, epithelium tends to spread from these bronchial openings over the internal surface of the cavity and in the course of time some chronic lung abscesses become lined with epithelium. An epithelialized cavity may then persist as a cystic space which is liable to recurrent attacks of infection.

Some adjacent bronchiectasis may develop as a result of the prolonged inflammation, and these damaged areas are liable to stagnation with infection and the production of cough and further sputum.

Treatment. A lung abscess usually presents as a pneumonitis, the true condition only being recognized by the offensive character of the sputum. In recent years this foul or "putrid" type of abscess has become uncommon and the diagnosis of the present, more benign, form is often only made as a result of radiography. Antibiotics, notably penicillin, used in high dosage in the early stages of the pneumonitis, can prevent the occurrence of the more severe symptoms in a number of cases. By the time the abscess has been diagnosed the bacteriology of the sputum is varied and only on occasions is a specific organism obtained on pure culture. In spite of persistent fever, high leucocyte count



(a) (b) (c) (d)

FIG 212 Lung abscess.

(a) Multiple lung abscesses in posterior segment of upper lobe and apex of right lower lobe.

(b), (c) Appearance after 2 weeks' intensive chemotherapy. Abscesses still show but inflammatory changes are reduced

(d) Appearance 4 months later showing complete resolution.

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Prolonged infection of bronchi will produce inflammatory changes in the bronchial wall and if there is undue infiltration and softening the walls will lose their rigid structure and will tend to distend either by traction or distension from coughing.

Some bronchial dilatations, particularly of a cystic form, are said to be of congenital origin but this theory is not adhered to at the present time, though there are occasional instances in which an anatomical deficiency in the wall of the bronchus could be pictured as leading to some form of bronchial dilatation.

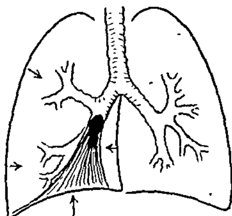


FIG 213 (*a*) Atelectasis Active collapse of a lobe exerts traction on surrounding tissues

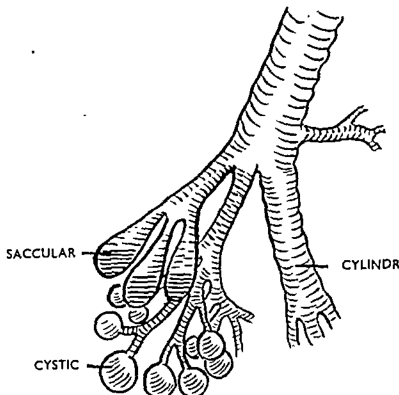


FIG. 213 (*b*) Types of bronchiectasis described according to their appearance and relationship to different sizes of bronchi.

Atelectasis plus infection is the most probable cause of clinical bronchiectasis and the process of events can be illustrated by the example of obstruction of a lobar bronchus. If there are enlarged glands which have narrowed the bronchus and an attack of bronchitis supervenes the œdematous mucous membrane and thick or sticky mucus may obstruct the already narrow tube. The air distal to the block is rapidly absorbed and the mucus which formed the block is sucked down into the finer bronchi where it is arrested again making the bronchial obstruction complete. If this mucus liquefies or is coughed up the lung will re-ærate, but in certain circumstances it persists and organizes so that the bronchial branches are completely and permanently cut off. The traction effect of the atelectatic tissue will not be marked on the more rigid bronchial walls, but in the more peripheral branches their less firm structure permits them to be distended and dilated. The nature and type of the dilatations depends largely on the site at which the obstruction occurs. If the obstruction occurs at the level of a second degree bronchus a cylindrical dilatation occurs, whereas at a third or fourth degree branch a saccular or glove-finger form is found and if the obstruction is close to the bronchioles then poorly supported

and toxæmia, chemotherapy should be persisted with and if penicillin is used it must be used in doses of at least two mega-units a day for at least 3 weeks. If, as a result of chemotherapy, the abscess remains static or even starts to diminish in size there is every hope that it will resolve spontaneously. Should it rupture into the bronchial tree the segment should be accurately localized and the patient placed in the appropriate position for postural gravity drainage. At the same time, if the cause is not obvious, the possibilities of neoplasm should be borne in mind. Steady reduction in size of the cavity over a period of weeks is normally observed and when all signs of toxæmia have ended, assessment of the local pulmonary damage is usually carried out by means of bronchography.

If the condition becomes chronic and a cystic space with or without bronchiectasis is present recurrence of infection is possible. This usually occurs after some months following an intercurrent infection. In such cases adequate chemotherapy is indicated and the whole position should be assessed to determine whether or not segmental excision is necessary. In young patients this would probably be the wisest course, but in more elderly the risks of surgery may be greater than the risks of recurrent infection. Prior to the introduction of penicillin many lung abscesses required external drainage, an operation which could only be performed through the adherent pleural cavity. The mortality of surgery was high and complications, in the form of secondary hæmorrhage and cerebral abscess, formidable, but there was no alternative. The picture has now completely changed and the only abscesses drained are those which are mistaken for localized empyemas. Healing, following drainage, may be complicated by persistent bronchial fistulæ.

Bronchiectasis

By definition bronchiectasis implies dilatation of the bronchi without any additional pathology, but in practice the clinical definition of the term implies dilatation plus signs and symptoms which are the result of infection. Accurate diagnosis can only be made following contrast filling of the bronchial tree and radiology—bronchography.

Ætiology. Two theories are advanced to explain the causation of bronchiectasis. One that it follows active collapse of lung or atelectasis, and the other that the walls of the bronchi dilate after they have been softened and weakened by inflammation. Bronchiectasis is commonly associated with lung suppuration but it also occurs in pulmonary tuberculosis and in connection with tumours, or any condition which distorts or compresses the bronchi.

If a bronchus is completely obstructed the trapped gases are rapidly absorbed into the pulmonary capillaries and what was an ærated sponge of tissue contracts into a solid mass of bronchi, vessels, and the connective tissue elements which represent the former alveolar structure. This concentrated mass exerts tension on all surrounding tissues and produces the following changes. Adjacent lung is over-expanded or stretched—compensatory emphysema; there is inward traction on the chest wall, the mediastinum is pulled towards the mass and the diaphragm is raised. When these traction forces have exerted their maximum pressure there is still one structure which they can affect, namely, the bronchi which have remained patent because of their rigid walls; these walls are gradually pulled on and will ultimately stretch. The whole area is functionless and any retained secretions remain stagnant, thus favouring infection which sooner or later will invade the area.

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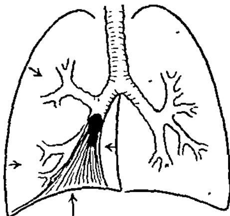


Fig 213 (a) Atelectasis. Active collapse of a lobe exerts traction on surrounding tissues

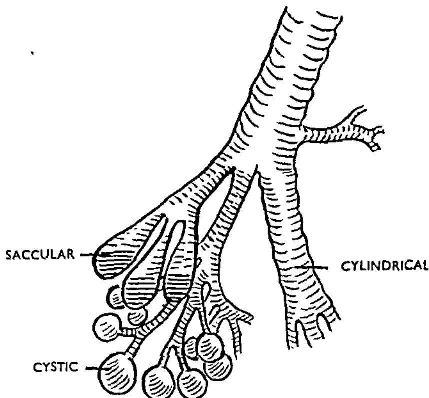


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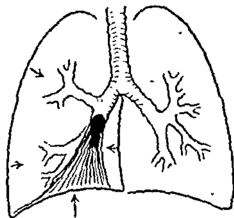


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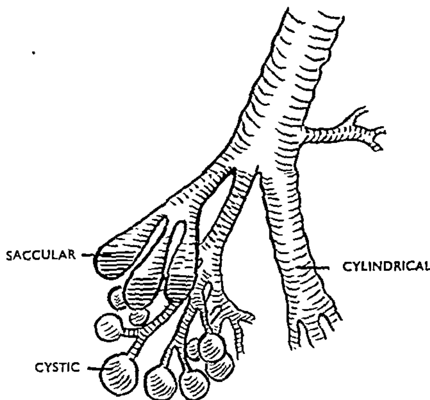


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If a bronchus is completely obstructed the trapped gases are rapidly absorbed into the pulmonary capillaries and what was an ærated sponge of tissue contracts into a solid mass of bronchi, vessels, and the connective tissue elements which represent the former alveolar structure. This concentrated mass exerts tension on all surrounding tissues and produces the following changes. Adjacent lung is over-expanded or stretched—compensatory emphysema; there is inward traction on the chest wall, the mediastinum is pulled towards the mass and the diaphragm is raised. When these traction forces have exerted their maximum pressure there is still one structure which they can affect, namely, the bronchi which have remained patent because of their rigid walls; these walls are gradually pulled on and will ultimately stretch. The whole area is functionless and any retained secretions remain stagnant, thus favouring infection which sooner or later will invade the area.

finding. In adults the most serious disadvantages are persistent cough and the offensive odour of the breath. Recurrent attacks of so-called pneumonia often characterize the history and are the result of pus retention in the diseased area; these respond quickly to antibiotics and do not cause the same inconvenience as a pneumonic attack though the physical signs are similar.

On examination the chest over the affected area is dull with restricted movement; there may be flattening of the chest wall and some displacement of the mediastinum. The breath sounds vary according to the state of the secretions in the lung and an important feature is that loud creaking or leathery râles can be heard, these changing in character after coughing.

Distribution. Excluding bronchiectatic changes that accompany tuberculosis the main site for infected bronchiectasis is at the base of the lungs. When the disease is localized the left lower lobe and lingular segment of the upper lobe are most commonly involved. Next in order of frequency comes the left lower lobe, then the right middle lobe, followed by the right middle and lower lobes. Bilateral lower lobe involvement and complete involvement of one lung is often seen, and a common pattern in many bilateral cases is disease affecting the left lower lobe and lingula plus the right middle lobe. Upper lobe bronchiectasis, as occurs in tuberculosis, is not so significant from the clinical point of view as these areas tend to drain spontaneously.

"Spill-over" infection into adjacent bronchi is sometimes seen in chronic cases and is due to the pus from the original site acting both as an obstructing and infecting agent.

Associated Factors. As has been indicated, upper respiratory tract infection is an important factor in the production and maintenance of bronchiectasis and if possible gross sinus infection should be eliminated before attention is turned to the lungs.

Investigations. The investigation and accurate diagnosis of bronchiectasis can only be made by bronchography. The instillation of radio-opaque oil or fluid undertaken with special care enables the bronchial tree to be outlined and the site of the dilatations identified. The oil is introduced into the trachea by dropping it directly through the cords or via a catheter passed through the mouth or nose. Puncture through the crico-thyroid membrane is another popular method. The thick oil is slowly introduced and trickles down the trachea into the bronchi during which time the patient is postured so as to guide it into individual lobar bronchi. When one lung has been adequately filled antero-posterior and lateral radiographs are taken. On most occasions it is desirable to do the two sides separately since bilateral filling will make interpretation of the lateral films almost impossible. Preliminary posture to remove all bronchial secretions should be undertaken in every case, and the efficiency of bronchography depends largely on the specialized technique that can be developed between the operator and the radiographer. Bronchography is not a procedure to be undertaken by the inexperienced, it will only have to be repeated. Lipiodol and neohydriol have enjoyed prolonged popularity as a contrast agent; newer substances such as dionysil are proving successful with the advantage that they disappear from the bronchial tree in a short space of time.

Treatment. The only radical treatment known for established bronchiectasis is surgical excision. Patients are usually assessed with this object in mind, but before any question of operation is considered a prolonged period of conservative and general treatment must be instituted. If excision is not considered practicable the patient must be prepared to continue with this conservative régime indefinitely.

tubes blow up into cyst or grape-like forms—a process which can occur in a short space of time.

Any form of bronchiectasis that remains uninfected can persist without symptoms for a number of years, but the probability of stagnation and added infection is considerable, and sooner or later inflammatory changes occur in the damaged area. Small, round-celled infiltration in the bronchial walls with inflammatory changes and fibrosis in the collapsed lung tissue lead to softening that exaggerates the bronchiectasis and produces symptoms. In many cases there is a steady source of infection from inhalation of infected particles from the naso-pharynx and sinuses; this not only maintains the infection but possibly causes further small areas of atelectasis.

As has been indicated, obstruction plays a large part in the causation of bronchiectasis and any condition which produces enlarged hilar glands may lead to kinking or actual pressure on the bronchial walls; whooping cough is a good example and glandular enlargement in tuberculosis is a common cause of bronchial occlusion. Similarly, foreign bodies and growths, such as carcinoma and adenoma, will lead to atelectasis with dilatation which is further exaggerated if infection follows. Glandular enlargement is responsible for lobar forms of the condition while in cases where the whole respiratory tract is catarrhal and infected the type of lesion will be multiple and patchy, as is seen in children with infected sinuses and chronic bronchial catarrh.

In the absence of infection symptoms are absent or negligible and the diagnosis may only be made by radiology. When, however, the dilated areas become infected there is an accumulation of pus which gives rise to continuous cough and purulent sputum, and if there is any active ulceration hæmorrhage may result. Apart from inflammatory changes round the bronchi the state of the bronchial mucosa is interesting. Areas of ulceration are uncommon and even in the presence of prolonged inflammation the bronchi are lined with a deeply staining, thick layer of epithelium which, though showing desquamation, does not show loss of continuity in the surface.

Signs and Symptoms. The signs and symptoms are almost entirely dependent on the degree of infection and the position of the disease. If the pus is able to drain by the action of gravity the symptoms are not generally severe, but in most cases where one or both lung bases are involved pus accumulates until it is coughed up. Cough is most noticeable in the morning and evening and may be exaggerated by alteration of position.

The amount of sputum varies from a trace to as much as half a pint a day. This measurement is not always a full record of the amount produced since a considerable quantity is swallowed. In children under the age of 4 almost all the sputum is swallowed and the amount can only be gauged by performing gastric lavage before breakfast. At the same time the amount of actual sputum apparently produced from the lung may be modified by oronasal sepsis. Pus trickling down from sinuses may be coughed up and add its bulk to the sputum. Bleeding is fairly common and in most chronic cases a history of hæmoptysis can be obtained, but the amount of blood lost is rarely excessive.

The general condition of the patient depends on the degree of toxæmia. Children are apt to be stunted and to have bad breath with a large, swollen, furred tongue, and spongy gums; their complexions are sallow and whereas they may lead ordinary lives the degree of toxæmia is sometimes only recognized by the improvement that results after the disease has been remedied. Clubbing of the fingers may occur but is by no means a constant

Surgical Treatment. Excision of the bronchiectatic area or areas will give in many cases complete relief. Lobectomy if the disease is confined to one lobe is a most satisfactory operation, giving 85–90 per cent cures with a negligible mortality. When, however, the disease affects more than one lobe the results are not so good, and in diffuse bilateral cases surgery may only be palliative, though a patient who is coughing up eight or ten ounces of sputum per day will be more than grateful to have this reduced to half an ounce or an ounce.

The amount of bronchiectatic lung that can be removed is considerable. The loss of a single lobe is rarely noticed, and many children grow up after a single lobectomy to take part in active athletics. The loss of two lobes does not cause much inconvenience, and a reasonable existence is possible if a whole lung is removed. Cases have been encountered in which the lower and middle lobes on both sides have been excised leaving the patient to exist on two upper lobes.

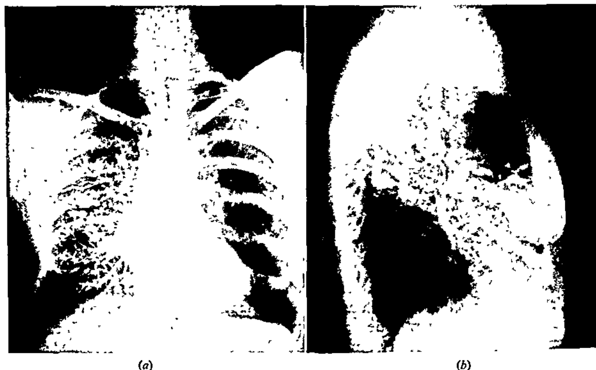
The most suitable and satisfactory time for operation is at an age when the child can co-operate in the preliminary preparation and after-treatment. Atelectasis following lobectomy is much more difficult to treat in an unco-operative, small child than in one who is old enough to help himself. The ideal age is probably between 12 and 18 years though many cases are undertaken at younger ages and lung resection is sometimes performed in the first 2 or 3 years of life. In adult life, if the disease is long-standing, the decision to operate becomes more difficult because the disease is rarely confined to one lobe. The indications have to be well defined if surgery is to be considered in a patient over 30 or 35 years of age.

Operation. Excision of any segment or lobe of lung is a straightforward and routine procedure. The basis of the operation consists in isolating the bronchus close to its parent stem and dividing it, and at the same time identifying and severing the corresponding branch of the pulmonary artery. The veins are also secured close to their main trunk. Division of the bronchus close to its main stem is an important step in the operation and here the bronchial arteries may require ligature. The open, cut end of the bronchus is sutured by interrupted, non-absorbable stitches to make an airtight closure which is reinforced by covering with adjacent tissue.

For the actual operation the patient is placed in the face-down position or in the lateral posture, and a long, postero-lateral incision passing below the angle of the scapula serves to expose the muscles of the chest wall. The latissimus dorsi muscle is completely divided and the back end of the trapezius and serratus magnus muscles are cut. The chest is entered through the fifth or sixth intercostal space, and the thin, membraneous pleura divided. The lung will then collapse and the incision is stretched open by powerful mechanical retractor. Adhesions of the lung to the chest wall are divided, many of them requiring hæmostasis during the process, and the affected lobe is freed from all adhesions. In most cases this is not a difficult process but if an empyema has been present it may be difficult, and the freeing of the lobe can only be effected after extrapleural stripping. At the root of the lung the black, and usually enlarged hilar glands, are visible and surrounding them or entering their substance are a number of small vessels which require ligature.

The first step is to identify the lobar bronchus and dissect it out. The distal end of the bronchus is then clamped and the bronchus cut across. The bronchus is trimmed and closed with interrupted sutures. Using the clamp on the distal bronchus as a retractor, the corresponding pulmonary artery is exposed and dissected until it is certain that the

Conservative Treatment. The basis of conservative or medical treatment is to establish drainage of the stagnant and infected areas by using postural drainage. This consists in placing the patient so that the affected sacs or dilatations are facing downwards and their contents can drain under gravity. Sepsis is thus eliminated and considerable improvement in symptoms and general condition of the patient follows. Antibiotics may also be used, but their prolonged use is not always considered wise and it is better to treat the patients by short, intermittent courses of suitable antibiotics. Postural drainage is of inestimable



(*a*) (*b*)

FIG 214 Right bronchogram.

Anteroposterior and lateral radiographs showing gross saccular changes in the middle lobe and anterolateral segment of the upper lobe. The lateral view is essential for accurate localization

value so long as it is carried out efficiently and thoroughly; its use for a few minutes night and morning is of little value, it must be used for several hours a day, and indeed, if the patient can be trained to sleep in the appropriate position drainage will occur during the night and will make it less necessary during waking hours.

As the disease commonly affects the bases, the inverted position is the one most often used. The patient lies on the face with the buttocks at a higher level than the shoulders; this can be achieved by the patient leaning over the edge of the bed and resting with the arms on the floor, or by lying over a wedge so that the lower limbs balance the upper part of the body. Wedges with the centre height 18 in. or 20 in. on a thin frame can be easily constructed and can be made to take to pieces in the patient's home. At the same time physiotherapy can play a valuable part in assisting postural drainage and expansion of the bases. In the case of postural drainage of the middle lobes the patient lies flat on the back with a slight inclination towards the infected side. There is nothing to prevent the patient being ambulatory during conservative treatment, and indeed, they may benefit by being able to take active exercises. In addition to local treatment, the nasal sinuses and throat must be examined for gross sepsis which, if present, should be treated.

affected side. Energetic coughing and posture may cause the obstructing mucus to drain away. For the first hour or so this should be tried, but if it fails, a strong case can be made out for bronchoscoping the patient in bed without much delay. This enables the mucus to be removed and the lobe to reventilate, thus relieving a dangerous condition in the space of a few minutes. Persistence of atelectasis may well result in bronchiectasis in addition to failure of the lung to re-expand and obliterate the pleural cavity.

Results. If excision of lung can be effectively carried out so that all bronchiectatic areas are removed there is every hope that cure will be complete. If diseased tissue is left behind there will inevitably be some cough and sputum following the operation, and in this lies the difficulty of selecting patients for operation. A clear cut bronchiectasis, localized to one lobe, with no infection in the upper respiratory tract, will almost certainly lead to success but the pattern of the disease is rarely as clear cut as this. More commonly two lobes, notably the left lower and lingular, will be involved and these may be associated with involvement of the right middle lobe. Bilateral operations are performed but are proportionately less successful than unilateral cases. The principal consideration of the surgeon is to decide whether or not he can obtain complete excision without endangering the patient's respiratory capacity, and if he cannot excise all the diseased lung he must be prepared to find persistence of symptoms, even though these are diminished, and the likelihood of slow spread of the disease. In cases where the symptoms are really severe and patients are coughing up to 10, 12, or even 20 ounces of pus a day, excision of the most damaged area will give a symptomatic relief that is welcomed by the patient.

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vessel identified is the one that supplies the affected lobe. Anteriorly, the pulmonary veins are identified and if an appropriate lobar or segmental bronchus can be recognized, it also is dissected clear and divided between ligatures. Theoretically this should leave a free lobe but in practice the fissures are rarely complete, and the interlobar fissure has to be deepened until the lobe can be removed. The remaining lobe, or lobes, are then re-expanded by the anæsthetist and inspected for any bleeding or leak of air. Minute pinpoint bubbles are not important but a persistent escape of air requires a stitch to close a bronchial opening. A closed tube is then inserted into the bottom of the pleural cavity and attached to a water-seal and the chest wall closed in layers.

Post-operative Treatment. Shock or gross disturbance is uncommon unless there has been excessive bleeding or much handling of the tissues, and in any case this should have been remedied by blood transfusion during the operation. The patient is returned to bed and is given oxygen until it is certain that his breathing is not laboured and he is not distressed. Pain can be controlled by a variety of drugs but it is not advisable to use heavy doses of morphia which would depress the cough reflex and destroy the ejecting mechanism of the lungs, namely, cough. The water-seal tube should show a free swing in the bottle with occasional bubbling of air and a collection of blood-stained fluid which in 24–48 hours will amount to 7–15 ounces. As soon as the blood pressure has risen the patient is gradually sat up and encouraged to breathe on the affected side. He will have been trained prior to the operation in breathing and in the necessity of coughing which keeps the bronchial tree free of mucus and loose secretions. In 24 or 48 hours the water-seal tube is removed, so long as radiography has shown the lung to be fully expanded and occupying the whole of the pleural cavity. When this has been done the patient can sit out of bed and be walking on the third or fourth post-operative day. In an uncomplicated case the patient should be fully dressed and walking about at the end of a week, and in 2–2½ weeks ready to return home.

Complications. The principal complications of lobectomy are atelectasis of the residual lobe or lobes, and broncho-pleural fistula. Until 3 or 4 years ago there was an appreciable incidence of *broncho-pleural fistula* occurring between the tenth and fourteenth post-operative day. The stitches closing the bronchial stump had either become infected or given way with the result that a free communication between the bronchial tree and the still open pleural cavity existed. Unless this was quickly remedied secondary infection and an empyema resulted. Since the introduction of antibiotics and the knowledge that immediate and prompt re-expansion of the lung was essential, the number of fistulæ has been reduced to a minimum. When they do occur they will be recognized by expectoration of blood-stained fluid or pus (the contents of the pleural space) and the persistence of this until the pleural cavity is fully obliterated. Should a fistula occur before the lung is expanded a potential total empyema occurs, but more usually the fistula drains into a small, posterior pleural pocket which can be obliterated by repeated aspiration and chemotherapy, and failing that by rib resection and drainage.

Atelectasis results from obstruction of a bronchus by sticky mucus or blood which has not been coughed up. This is most likely to occur on the operation side where pain inhibits free movement of the chest, and in consequence the obstructed lobe becomes airless and atelectatic. The onset of this condition can readily be recognized by change in the patient's appearance; he becomes breathless, cyanosed, and distressed usually with wet or bubbly breathing, and on examination the trachea will be found drawn towards the

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lung cancer existed, and this was confirmed by Wynder and Graham (1950) in the United States. The latter authors have shown that tumours can be produced in mice by using a condensed extract of cigarettes, and a carcinogen 3:4—Benzpyrene has been suggested as a possible responsible agent by Stocks and Campbell (1955). In addition to cigarette smoking, there is no doubt as to the higher incidence of death rate in town than in country, and in town the proximity to gas works or smoky atmosphere seems

REGISTRAR GENERAL'S REPORT FOR 1954

DEATHS

	1944	1948	1952	1954
Tuberculosis (Respiratory) .	19,815	18,798	9,335	7,069
Cancer of Stomach . .	13,155	14,392	14,409	14,114
Cancer of Large Gut . .	10,150	10,438	10,822	9,528
Cancer of Breast . . .	7,366	7,907	8,344	8,441
Cancer of Œsophagus . .	2,196	2,311	2,325	2,236
CANCER OF LUNG . .	6,568	10,162	15,218	16,331*

* This figure consists of 13,995 males and 2,336 females. 10,695 of these deaths occurred between the ages of 50 and 70.

to be established. In short, it would appear that the urban dweller who is a heavy cigarette smoker over a long period is far more at risk than the countryman who is a non-smoker.

Histology of Lung Cancer. Conventionally three forms of histological type have been recognized:

(1) Squamous carcinoma.
(2) Adenocarcinoma.
(3) Oat-cell.

Differentiation is of important clinical and prognostic significance since tumours behave differently and some authorities have added to the three groups a fourth, or even fifth type. Walter and Pryce (1955), after an analysis of a large number of necropsy and operation specimens, suggest the standard classification with the addition of a polygonal-cell type and also the invasive type of adenoma which is ordinarily non-metastasizing. Following their classification, the oat-cell tumour is regarded as an entity, the cells being

small with round or oval nuclei arising from the bronchial epithelium. In a proportion of the cases the histological picture was not uniform and rosette and tubule formation was observed making it difficult to differentiate them from adenocarcinoma.

(1) THE OAT-CELL TYPE tends to form early and to produce massive regional metastases with the result that this tumour has often been referred to as a hilar tumour. The glandular involvement has a serious effect on the prognosis.

(2) SQUAMOUS-CELL CARCINOMA shows keratinization and no indication of tubule formation; the cells are of squamous type and character, tending to arise in large bronchi which they obstruct at an early stage in their growth.

(3) ADENOCARCINOMA shows evidence of glandular structure with high columnar type of epithelium and the formation of tubules and mucus production. These tumours are often peripheral and rounded.

(4) POLYGONAL-CELL CARCINOMA shows no evidence of glandular or squamous differentiation and is probably an anaplastic variation of glandular forms rather than being connected with the oat-cell type.

The necropsy series of Walter and Pryce show that squamous-cell types are present in 20 per cent, adenocarcinoma in 28 per cent, oat-cell in 37 per cent, while in the surgical or operation series squamous-cell tumours account for 60 per cent of the total while adeno- and oat-cell carcinoma each give figures of 15–20 per cent. These figures show that the oat-cell, which was previously regarded as being the most common, only accounts for just over one-third of the total tumours. The same authors, relating the site of origin to the histological pattern, show that adenocarcinomas are always peripheral, a feature that they share with polygonal-cell growths whilst squamous-cell tumours are more often central than peripheral, with oat-cell carcinoma being indeterminate as regards site. The term peripheral, as used, refers to tumours arising in minute bronchi or bronchioles. It is possible that a tumour which is histologically peripheral may lie close to the hilum in the true anatomical sense.

Symptomatology. The signs and symptoms of pulmonary cancer are immensely varied, depending first on the presence of distant metastases, and secondly on the site of the primary tumour and its relation to other structures. In general, distant or blood-borne metastases are not as common as might be expected when the enormous area of the vascular field of the lung is considered. Many tumours undoubtedly tend to spread into or along the walls of the veins, whereas others metastasize along the lymphatic channels. Apart from secondary glands within the neck, the common sites for distant metastases, in approximate order, are: brain, bone, liver, adrenals, and skin.

For convenience, the symptomatology can be divided into three groups:

(1) tumours lying close to the pleura;

(2) tumours within the lung substance; and

(3) tumours lying against the mediastinum.

(1) PLEURAL TUMOURS. A growth on the actual surface of the lung readily becomes adherent to the parietal pleura and tends to spread along this structure without necessarily transversing it, or giving rise to seedlings in the pleural cavity. Both these latter events can and do occur, but there is a definite tendency for the pleural aspect of the tumour to remain localized to its parietal element. Pain and heaviness over the adhesion and the presence of a clear effusion are common. A frankly blood-stained effusion results from multiple deposits which ulcerate and give rise to bleeding. On the other hand, it is

important not to regard every blood-stained effusion as being neoplastic in origin unless it is certain that the first aspiration is definitely blood-stained.

(2) TUMOURS WITHIN THE LUNG. Within the main bulk of the lung tissue a growth can silently expand over a period of months without causing any secondary changes. Bronchi may be pushed to one side and the size of a tumour up to the dimensions of a tangerine may be accommodated without incident. On the other hand, as most tumours arise from a bronchus, secondary signs of bronchial obstruction may be expected. Hæmoptysis will occur from ulceration of the tumour and a variety of secondary changes may result from partial or complete obstruction of a large bronchus. Atelectasis is common and if infection supervenes pneumonitis, bronchiectasis, and lung abscess may all be encountered, and the secondary changes from these complications may be the initial ones noticed by the patient. A breaking-down growth usually shows an eccentric abscess with irregular walls and some of the peripheral type are associated with hypertrophic pulmonary osteo-arthropathy or with neurological changes, both of which are improved if the tumour can be removed.

(3) MEDIASTINAL TUMOURS. Any structure within the mediastinum may be involved by primary tumour or secondary glands. The superior vena cava can be obstructed when it is usual for the phrenic nerve to be involved at the same time; the vagus and recurrent laryngeal nerve, if paralysed, will lead to loss of voice, and lower down a growth invading the pericardium may lead to effusion. If infiltration occurs along the main pulmonary veins into the auricle, auricular fibrillation may occur. Pressure on the œsophagus by mediastinal glands will produce dysphagia.

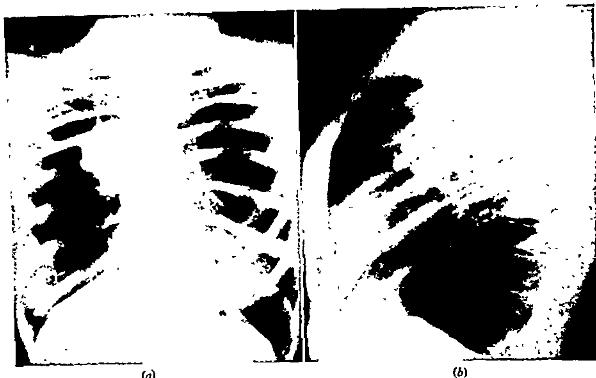
General Features. In the absence of bronchial obstruction or suppuration, the general symptoms are vague. There may be loss of weight and asthenia but in many cases a period of observation and symptomatic treatment results in the patient improving and putting on an appreciable amount of weight, thus giving a misleading impression as to the true origin of the condition. The whole picture can be modified by the presence of secondary deposits; the most misleading of these occur in the brain when a secondary in the frontal lobe may give practically no symptoms until the condition is well advanced. Secondaries in the adrenals may lead to marked wasting and asthenia—a classical picture of Addison's disease. When suppuration occurs in the lung the picture may be that of pneumonitis or bronchiectasis, with cough, sputum, and toxæmia dominating the condition.

Diagnosis. It is virtually impossible to diagnose cancer of the lung on clinical grounds. The silence of onset and the comparatively slow evolution of many tumours make it necessary to depend on other methods—namely *radiology*. At the least suspicion of any abnormal feature in the chest a radiograph should be obtained and if any abnormal shadow is present this should be investigated in extenso until a diagnosis is made. Many shadows of inflammatory origin may be seen to resolve with repeated radiographs, but if resolution is incomplete the suspicion must remain and investigations should be carried out even to the extent of thoracotomy.

Bronchoscopy is the most valuable diagnostic measure. An intrabronchial growth can be visualized and biopsy taken. Mobility of the bronchi can be observed and if there is any external pressure from enlarged glands it can be noted. Bronchoscopy is a simple and routine procedure at the present time and can be performed with a minimum of disturbance under local or general anæsthesia. After radiography it is the most important investigation.

Bronchography plays a useful part in diagnosis when the growth lies beyond the range of the bronchoscope. The site of the obstruction can be visualized and secondary changes such as bronchiectasis outlined.

Investigation of the sputum for malignant cells is a method which in expert hands has considerable accuracy, but a positive finding may only be made after repeated examinations.



(a) (b)

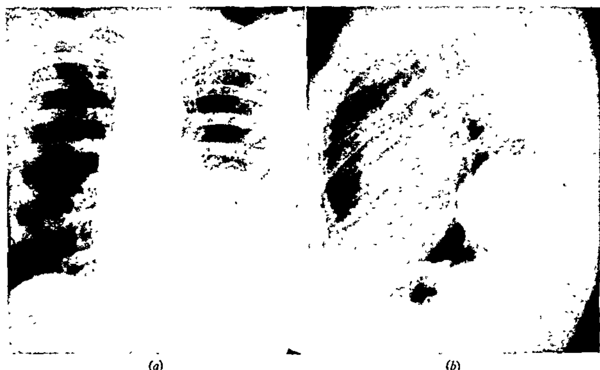
Fig 215 (a), (b) Oat-cell carcinoma of right upper lobe. Massive involvement of glands.

Finally, there is *thoracotomy* which is indicated when doubt still exists and other methods have given negative findings. It is unusual for a thoracotomy to be carried out without discovering an early tumour.

Life-history. Cancer of the lung is often a slow-growing tumour and cases have been recorded in which an untreated patient has lived 3–4 years. The average expectation of life following diagnosis without treatment is, however, 3–6 months. The approximate age of the tumour prior to diagnosis is 6–9 months, giving a total of 9–15 months. Unless some fortuitous circumstance, such as hæmoptysis or routine radiography, brings the case to notice, the average patient wastes nearly 6 months from the time of his first, insidious symptoms until he reaches the surgeon. Much of this loss of time is the responsibility of the medical profession who are not sufficiently aware of the possibility of cancer and who treat the symptoms palliatively, or are slow in making a diagnosis.

The highest incidence of lung cancer is in middle-aged men, but no age is immune and cases in which a growth has developed in young men and women in the twenties are not uncommon. Comparable with other forms of malignant disease the prognosis is worse in the younger age groups.

Treatment. The only hope of long-term survival lies in excisional surgery. Radiotherapy has a part to play but its effect is rarely curative and should not be used in



(a) (b)



(c) (d)

FIG. 216 Carcinoma of lung

(a), (b) Large rounded shadow in left lower lobe No involvement of glands.
(c) Tomograph showing irregular density in small squamous cell carcinoma in lower lobe
(d) The large roundish tumour in upper half of right lung proved to be an adeno-carcinoma which was treated by pneumonectomy.



FIG. 217. Carcinoma of lung.

(a) A triangular opacity at right base partly due to growth and partly due to atelectasis.
(b) Appearance immediately following pneumonectomy.
(c) Appearance of chest two months after pneumonectomy showing traction of mediastinum and flattening of the right chest. The remainder of the space is filled in with fibrin and effusion.

preference to surgery. It is regrettable that the proportion of cases of patients suitable for surgery is small when the whole bulk of lung cancer is considered. An actual figure of operability cannot be readily obtained and there are wide variations in the reported series. Possibly not more than 15–20 per cent come to surgery.

The collected figures from the Birmingham Hospitals (1954) afford a guide as to the present position in regard to treatment. Out of over 4,000 cases 52 per cent were not treated, 28 per cent were given radiotherapy and 20 per cent came to surgery. In this latter group resection was carried out in 12 per cent.

The fate of the untreated 52 per cent showed that only 5 per cent were alive at the end of 1 year and 1 per cent at the end of 2 years. Radiotherapy (28 per cent of the total) showed 13 per cent and 3 per cent survivals at the end of 1 and 2 years respectively. The resection cases had 48 per cent alive at the end of the first year and 34 per cent at the end of the second.

The problems that face surgery lie under two headings, the first concerned with the situation of the growth, and the second with the general condition of the patient. Exclusion of distant metastases is a sine qua non and in considering the general condition, assessment of respiratory function and the ability to withstand major surgery come into the picture. Patients with rigid emphysematous chests and a tendency to bronchospasm are not good surgical risks, and these factors, coupled with the mechanical possibility of removing growth and regional glands, are the main points in the selection of suitable candidates for operation.

The standard operation for carcinoma of the lung is *radical pneumonectomy*, a *removal of the lung by dissection of its hilar elements so as to include the total gland field*. The residual hemithorax slowly fills with effusion and fibrin at the same time as retraction of the mediastinum and chest wall occur. Complete obliteration takes several months. In some instances when the tumour is localized, or in which the condition of the patient would not withstand pneumonectomy, lobectomy is a practical proposition; indeed, the results of more limited operation are paradoxically nearly as good as those for pneumonectomy, but the answer to this lies in the necessarily more rigid selection of patients.

Operation (Pneumonectomy). Pneumonectomy is carried out through a long thoracotomy incision in which the fifth or sixth rib is removed. The lung is freed from adhesions and if the lung is adherent to the chest wall the dissection is carried out extrapleurally. The mediastinal tissues are opened and on the right side the paratracheal glands and those lying in the anterior mediastinum are dissected downwards. On the left the gland field anterior to and under the arch of the aorta is removed. The main bronchus is then freed and divided as close to the carina as possible and its central end closed with a series of fine, interrupted sutures. The dissection next includes the glands of the bifurcation and those lying on the œsophagus and posterior pericardium. With an extensive lesion the pericardium is opened to expose the pulmonary veins and main pulmonary artery. The artery is secured by firm ligature or sutures and the inferior and superior pulmonary veins tied and divided. Anteriorly any glandular tissues associated with the lung root or close to the phrenic nerve are widely excised and the lung removed. The bronchial stump is then covered by adjacent tissue divided from pericardial edges or œsophageal fascia, or by a pedicled intercostal muscle graft. The chest wall is then closed without drainage.

In most cases there is surprisingly little loss of blood and little evidence of shock. In the absence of complications the patient may feel fit enough to leave his bed in 1 or 2 days and to start moving about. In 10 days the average patient should be freely ambulant and fit to go home in a fortnight or 3 weeks.

The main complications after the operation are concerned with the behaviour of the bronchial stump and the condition of remaining lung. In the early days of pneumon-

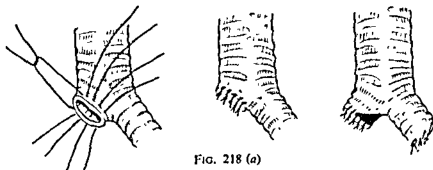


FIG. 218 (*a*)

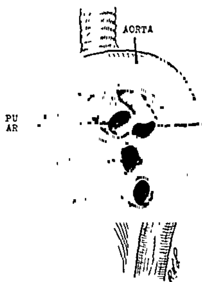


FIG. 218 (*b*)

(*a*) Method of closing the bronchus The stump is cut almost flush with the parent stem The diagram on the right shows that if a long stump is left it will act as a "sump" for infection.
(*b*) Diagram of the relation of the hilar structures on the left side

ectomy bronchial fistula was a common finding. If it occurred pleural infection followed, and this required drainage and later thoracoplasty to close the dead space. At the present time fistula formation is rare and complications on the non-operative side uncommon. The important part in the after-treatment is to ensure that the remaining lung stays free of secretions. This can be helped by coughing, breathing exercises and postural drainage. In the presence of bronchospasm or atelectasis the respiratory function of the patient is in danger and most of the fatalities result from infection in the residual lung.

Results. Recently several series of the results of a large number of resection cases have been published by Bignall and Moon (1955), Price Thomas (1952), Sellors (1955), and others. There is considerable measure of agreement in the figures which may be taken as representative of current practice.

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Arteriovenous fistula of the lung may take a variety of forms. Its characteristic feature is cyanosis and clubbing due to vascular shunt, which gives an audible murmur. Ligature of the vessels leading to the angiomatous area may avoid excision of the affected area of lung, but the important thing is to remove the shunt. The condition may be multiple and associated with angiomas elsewhere.

Tumours of the Mediastinum. Mediastinal tumour is a term that can be used to include fluid-filled cysts and solid masses which arise in or close to the mediastinum. These growths, which can only be recognized radiologically, afford considerable diagnostic



FIG 219 Adenoma of bronchus

Rounded shadow of right chest lies close to the mediastinum. This shadow had been observed for 15 years and had recently increased in size. It later completely obstructed the lower and middle lobe bronchi The tumour was removed by middle and lower lobe lobectomy

problems but as the majority of them steadily progress in size their early removal is advisable. Frequently it is only after excision that their true nature is appreciated. Rounded tumours lying in the anterior mediastinum are most commonly retrosternal goitres, thymic tumours, dermoids, or teratomas; a posterior mass lying close to the vertebral column is most likely to be of neurogenic origin, and radiological shadows close to the pericardium or œsophagus are likely to be pericardial cysts or cysts of enterogenous character.

Adenoma of the thyroid if lying within the mediastinum tends to displace the trachea and give rise to breathlessness. It may also obstruct the great veins while hæmorrhage into its substance is an occasional cause of asphyxia. Rare cases are seen in which an ectopic goitre lies low down in the posterior mediastinum. Nearly all retrosternal goitres

The operative mortality in the early 1940's was in the neighbourhood of 20 per cent, but during the last few years this has been more than halved. A further 12–15 per cent died before 6 months had elapsed but at the end of 2 years 35–40 per cent of the original total were alive. During the next 3 years there was a slow fall in the survival percentage and at the end of the 5 year period 20–25 per cent were still alive. The trend of the more recent resection figures suggests that the 5 year survival period would be 4 per cent or 5 per cent higher. Belcher (1956) and his colleagues reporting on 264 lobectomy cases with a 4 per cent mortality showed that the probable 5 year survival rate was something of the same order. If the operation mortality rate is excluded a patient treated for a carcinoma of lung by pneumonectomy, or where practicable by lobectomy, would at the present time have a 28–32 per cent chance of living for at least 5 years.

The presence of glandular metastases at the time of operation has a very sinister bearing on the prognosis. 80–90 per cent of those with involved regional glands did not survive the 2 year period; this emphasizes the very reasonable chances of success in operating on early cases without glandular involvement.

Benign Tumours of Lung

Adenoma. The most common benign tumour of lung is the so-called adenoma of the bronchus; this tumour, whose origin has caused considerable discussion, has many points in common with mixed salivery tumours, and is said to arise from the mucous glands in the wall of bronchus. It presents as a dumb-bell with a berry-like projection into the bronchial lumen and a deeper part extending into the peribronchial tissues. Its pathological behaviour is interesting in that these tumours while showing active local invasive characters do not give rise to metastases. Exceptionally, one of these tumours which histologically resembles adenocarcinoma does produce glandular deposits.

The symptoms depend largely upon the degree to which the bronchus in which they arise is obstructed. The intrabronchial part of the adenoma can give rise to hæmoptysis and if it completely occludes the bronchus it will produce atelectasis which, if associated with infection, proceeds to bronchiectasis or abscess formation. The clinical picture, therefore, is dominated by the secondary changes in the lung. Radiologically, the tumour appears as a more or less rounded mass usually in association with a bronchus of moderately large size. Bronchoscopy may reveal a mass projecting into the lumen and a bronchogram may demonstrate a square cut or convex filling defect quite unlike the "rat-tail" appearance seen in most cases of lung cancer. Even though these tumours are regarded as relatively benign, their tendency to grow and the occasional risk of malignant change makes it imperative that they should be excised. The most common operation performed is a lobectomy but if a tumour arises in a main bronchus it may be possible to remove it by excising a sleeve of that bronchus and restoring continuity by end-to-end suture, thus preserving lung tissue which, if healthy, would otherwise have to be sacrificed.

Hamartoma. This type of tumour is a pathological rarity and there are conflicting views as to its nature. Histologically it consists of all the elements that may be found in the structure of a bronchus with occasional ossification of cartilage. These tumours are not usually completely rounded and some resemble early carcinoma; they should be excised.

Fibroma of lung has a particular interest in that it may be associated with gross clubbing of the fingers and toes as well as cyanosis. It presents as an oval or rounded shadow, usually close to the mediastinum.



FIG 221. Large anteriorly placed teratoma which was successfully removed.

(a), (b) A shadow extends across the mediastinum to both lung fields The condition was benign
(c) Radiograph of the specimen removed from the above, showing cystic character of the tumour with areas of ossification and tooth formation.

can be removed from the neck but if a further exposure is needed, a sternal splitting incision exposes the anterior mediastinum and gives freer access.

Thymus tumours are of extremely complex pathological origin and may appear as small, rounded masses barely visible radiologically or as large tumours with little apparent connection with the thymus gland. Some cases are associated with myasthenia gravis, as a result of which their presence is first recognized. Drew Thompson (1955) has put forward the suggestion that Hodgkin's disease is in effect a tumour of the thymus gland itself; many cases of mediastinal tumours suspected to be Hodgkin's or lymphosarcoma

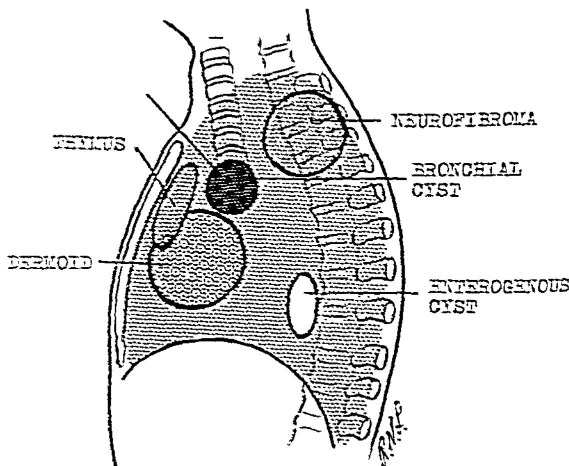


FIG. 220. Diagram to illustrate the sites of the more common forms of mediastinal tumour.

have proved to be thymic in origin. For a more detailed account of these tumours see Chapter IX.

Dermoids and Teratomas. The upper mediastinum is a common site for dermoid cysts or teratomas. They arise close to the arch of the aorta and expand into either lung field, sometimes attaining a large size. These tumours may be cystic, containing sebaceous matter and hairs, or semi-solid and loculated without ectodermal contents. Malignant teratomas respond to deep X-ray therapy whereas the dermoid cyst does not. The differential diagnosis is often complicated and these masses can easily be confused with aneurysms. Excision is the treatment of choice but on occasions when a large cyst is present the removal may be incomplete, in which case the edges of the cyst are brought to the chest wall (marsupialization), and the residual cyst lining can be destroyed later by caustics.

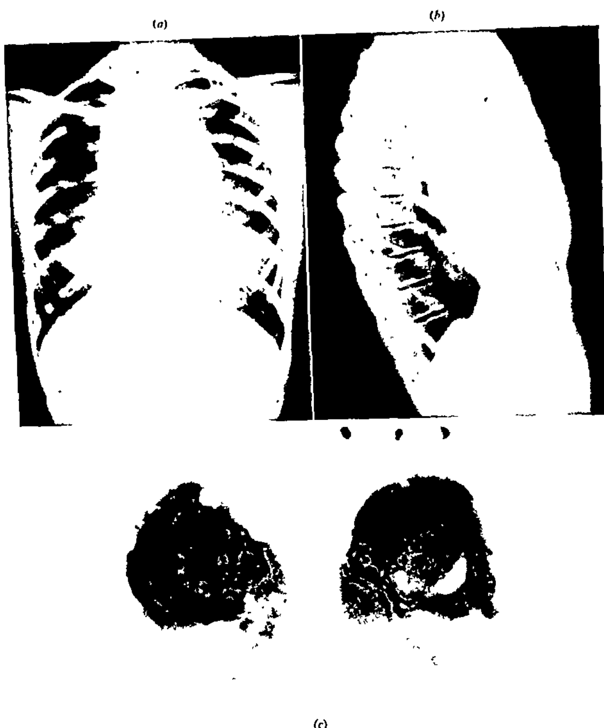


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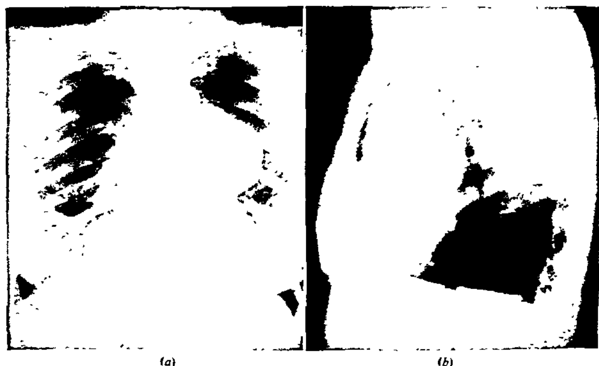


FIG. 222. Large dermoid cyst.
Arising from the anterior mediastinum and projecting into the left lung field.

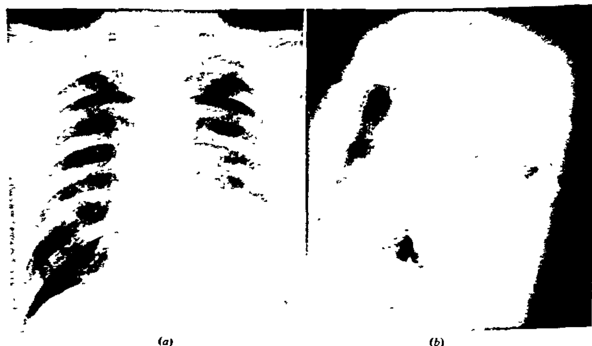


FIG. 223. Neurofibroma.
Large rounded tumour at the left base posteriorly against vertebral column.

Neurofibroma is a term which includes tumours of neurogenic origin. The majority arise from the sheath of Schwann and they lie close to the intervertebral foramen and may arise within the canal or even within the spinal cord. They are usually clear-cut and rounded in outline and if of any size they erode the posterior parts of the ribs and may even produce spontaneous fracture. Occasionally they undergo malignant change and at any time may give rise to considerable amount of pain. Their excision is not a difficult matter unless there is widening of the intervertebral canal when a spinal projection of the tumour may be expected. On a few occasions a simultaneous laminectomy and thoracotomy have had to be performed for complete removal.

Bronchogenic cysts and enterogenous cysts are congenital malformations arising from the foregut and are closely related to the bronchus or œsophagus. These cysts are frequently lined by epithelium and may contain altered blood. Some enterogenous cysts have a peculiar interest in that gastric and even pancreatic elements may be found in their lining. Complete excision is usually possible though adhesions or even small openings into the bronchus or œsophagus may be present.

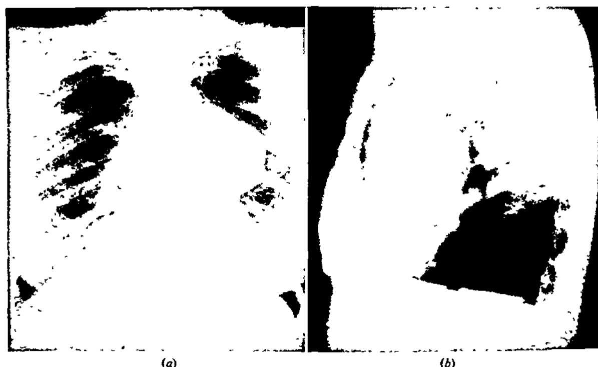
Pericardial cysts lie in close association to the main sac and are translucent, thin-walled structures containing clear fluid, hence the term *spring-water* that is often applied to them. There may be a communication between the cyst and the pericardium. Superficial pleural cysts of similar formation are occasionally seen.

Lipoma. A few cases have been recorded of massive, mediastinal lipoma whose lobulation is the characteristic feature. Occasionally a smaller form of lipoma occurs in relation to the extra-pericardial fat.

In addition to the above, there are a host of rare tumours and radiological shadows that can easily be confused with these masses. The difficulty of diagnosis has already been stressed, and with anterior mediastinal tumours in particular it is often wise to give a therapeutic dose of X-rays and to observe the response at the end of 4–6 weeks. Failure of the tumour to reduce in size is suggestive of its benign origin. If a diagnosis cannot be made it is justifiable in some cases to explore the mediastinal tumour and establish the true nature of the mass by biopsy if removal is not possible.

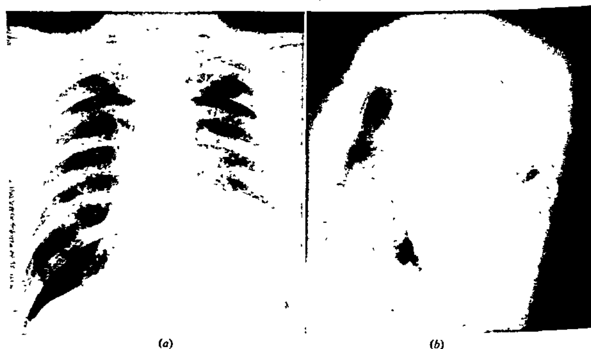
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(a) (b)

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Arising from the anterior mediastinum and projecting into the left lung field



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CHAPTER IX

SURGERY OF THE THYMUS IN RELATION TO MYASTHENIA GRAVIS

J. E. Piercy

History

Myasthenia Gravis was described in the seventeenth century but it was not recognized as a separate disease until 1900. Its association with epithelial tumours of the thymus had long been known but it was not until 1936 that Blalock at Baltimore removed such a tumour which resulted in a cure of the myasthenia gravis. In 1941 he gained a similar result by removing an apparently normal thymus gland from a myasthenic patient and reported on six further thymectomies by the end of that year. It was at that time that a thymectomy was first undertaken in this country at New End Thyroid Clinic by Sir Geoffrey Keynes, to be followed by a series of 280 cases.



Fig 224. Thymus gland removed from a patient aged 30 years suffering from myasthenia gravis.

Cause of Myasthenia Gravis

Myasthenia gravis is characterized by an abnormally rapid exhaustion of the voluntary muscles. It has recently been shown that the thymus gland may release or be intimately concerned with the release of a substance which affects neuromuscular transmission (Wilson, A.). This effect could be caused by the substance inhibiting the production or action of acetylcholine and thereby giving rise to rapid exhaustion of muscle. This secretion, extracted from thymus glands removed from patients suffering from myasthenia gravis, was shown to have a specific action on muscle nerve preparations reducing their contractions and also when injected into small animals giving rise to a temporary paralysis.

The manufacture and discharge of the causative substance by the thymus could explain the occasional remissions and fluctuations seen in myasthenia gravis and the fact that thymectomy can alleviate control or cure the condition. If, however, the disease has been present for many years, irreversible changes may have occurred in the muscles which would result in a poor or lessened response to neostigmine and to an unsatisfactory result from thymectomy.

The thymus gland increases in size from birth to puberty from which time it slowly diminishes in size to become atrophic in the aged although it does not disappear. It weighs anything between 5 and 40 grammes, varying considerably in normal individuals of the same age.

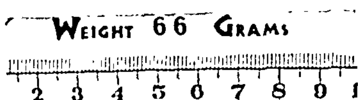




FIG. 225. A tumour in the left lobe of the thymus.

The Thymus of Myasthenia Gravis (Fig. 224). The thymus is not necessarily increased in size when associated with myasthenia gravis, nor does its size bear any relation to the severity of the disease. Its naked eye appearance therefore is not unlike that of a normal thymus and it should not be described as being either persistent or enlarged.

Microscopically, the thymus of myasthenia gravis is unlike the normal thymus and has

germinal centre or lymphoid follicle formations and increased cellularity of cortex and medulla.

A *thymoma* (Fig. 225) was found in 12–14 per cent of patients suffering from myasthenia gravis who underwent thymectomy. It is very probable that this type of neoplasm occurs only in myasthenia gravis and could be labelled thymo-lympho-epithelioma, being a mixture of epithelial and lymphocytic cells in varying proportions. The tumour, usually encapsulated and slowly growing, has all the characteristics of a benign neoplasm, although it must be considered potentially malignant as a small percentage of them may infiltrate the pleura and lung and more rarely give rise to secondary deposits within the thoracic cavity.



FIG. 226. Showing bilateral ptosis. An attempt is being made to raise the lids.

Incidence. Myasthenia gravis is more common in the female, in the ratio of 2:1, its highest incidence being between the ages of 15 and 40 years.

When associated with thymic neoplasm; however, it is relatively more common in the male. In either sex the majority with tumours occur between the ages of 30 and 55 years.

History, Symptoms and Signs

The early symptoms of myasthenia gravis are variable and are often dependent on the occupation of the patient. The shorthand typist would notice fatiguability of wrists and fingers, whereas the delivery boy weakness of his thigh muscles on bicycling. The clinical picture is never the same in any two patients, as any muscle or group of muscles may be first involved, slowly spreading to other groups with yet other symptoms arising. As there is an absence of neurological signs and of muscle wasting, an erroneous diagnosis of a functional state is often made.

The patient may first complain of drooping eyelids and double vision (Fig. 226), soon to be followed by difficulty in smiling and swallowing. Upper and lower limbs and trunk may next in turn be involved, speech becoming slurred and nasal, often associated with difficulty in swallowing and nasal regurgitation of fluids. The spread of fatiguability may advance quickly into a complete and generalized picture of the disease, or more slowly perhaps leaving the lower limbs uninvolved for many months. The process may be continuous or intermittent and associated with a series of relapses or remissions. *The Bulbar type* of myasthenia gravis is that in which only the muscles supplied by the bulbar nerves are involved, in particular those of the eyes and face.

Commoner Signs of Myasthenia Gravis. (All weakness increased by repetition of movements.)

(1) Ptosis of eyelids, occasionally unilateral; weakness in clenching eyelids.
(2) Limitation of movements of eyeballs, giving rise to double vision.
(3) Weakness of facial muscles (*a*) in showing teeth, (*b*) in smiling (snarling smile).
(4) Weakness in chewing; a drooping lower jaw.

(5) Non-explosive cough.

(6) Slurring of voice, which may become unintelligible after talking or reading aloud.

(7) Inability to extend fingers after hand is first clenched and dorsiflexed. If extension of fingers is possible, test strength to maintain this position.

(8) Inability to raise and maintain arms in partly raised position.

(9) Inability to raise extended leg when recumbent.

(10) Inability to dorsiflex foot against resistance.

(11) Inability of patient to sit up from a recumbent position with and without use of arms.

Many other signs and symptoms may be present such as aphonia due to weakness of the adductor muscles or stridor due to weakness of the abductor muscles of the vocal cords. The picture may be that of simple ptosis of an eyelid, bulbar or partial myasthenia or any degree of severity of the generalized state. The severe generalized picture of myasthenia gravis is a grave and pathetic one. The patient lies helpless in bed unable to speak, to swallow or to raise her limbs from the bed. An attempt at drinking causes coughing, dyspnœa and regurgitation of fluids through the nose. The mouth sags open and the patient is unable to cough up the outpouring of bronchial secretions.

Establishment of Diagnosis

(1) It is necessary to ensure that the weakness is a fatiguability of muscles which is increased by repetitive exercise and in the main improved by rest.

(2) Response without side effects to anticholinesterase drugs (neostigmine). The weakened muscles in myasthenia gravis respond to an intramuscular injection of 1–2 mgm. neostigmine. Some response will always take place, the increased scale of activity to normal or near normal taking anything from 10–30 minutes.

(3) The intravenous injection of tensilon rapidly gives rise to a brief increase of muscular strength in myasthenia gravis but to no response in other conditions.

(4) The presence of resistance to the depolarizing activity of decamethonium (C 10) may be used as a diagnostic test in occasional difficult cases of localized myasthenia gravis (Churchill-Davidson).

Treatment

The only medical treatment available in myasthenia gravis is by the anticholinesterase drugs; they do not directly influence the disease process and are not in any way concerned with the development of remissions They are used not in the hope of gaining a cure but temporarily to increase the muscular power and scale of activity, and are used with or without thymectomy. The drugs are repeated at intervals as the muscular power diminishes.

These valuable cholinergic drugs are antagonistic to cholinesterase and by diminishing its power allows for a fuller action of acetylcholine and, therefore, a stronger contraction of muscle.

Prostigmin or Neostigmine is a synthetic compound closely allied to physostigmin. It is made up in 15 mgm. tablets, one of which is equal in action to half a milligram by intramuscular injection.

Mestinon (pyridostigmin), a more recent drug, is an analogue of neostigmine, tablets of 60 mgm. being equal in strength to 15 mgm. of neostigmine. In the majority of cases of

myasthenia gravis it is longer acting than is neostigmine and gives rise to fewer side effects.

Overdoses of the cholinergic drugs give rise to stimulation of the parasympathetic, leading to uncomfortable side effects. The early and most common side effects are gastro-intestinal, being nausea, colic and vomiting associated with sweating, pallor and fasciculations of the voluntary muscles. It is often necessary that food be taken just before or with the drug, which will lessen or stop the side effects from occurring. Every patient must be studied individually as to the requirements of neostigmine. The aim is to gain as high a scale of activity as is possible. If the myasthenia is of a mild degree, one tablet, i.e. 15 mgm. of neostigmine, three or four times a day may suffice. Any degree of severity may be encountered, requiring up to forty or fifty 15 mgm. tablets each day, or even more. In the severe cases the tablets may need to be reinforced by one or two milligrams by injection. This could be necessary first thing in the morning if the patient were too weak to swallow tablets.

The decision in regard to the optimum dose of the cholinergic drug is made by trial and error, and no-one is better able to judge the requirements than the patient herself. The more severely myasthenic the patient is, the higher the dosage required.

The severity of the condition is often progressive, requiring the administration of an ever increasing dosage of the cholinergic drugs. In long standing cases the dosage required is often very large, as the condition may have become partially irreversible. In some of these patients, side effects, as shown by an intense stimulation of the gastro-intestinal tract, make it difficult to gain a therapeutic response unless atropine is given at regular intervals.

Indications for Operation. All patients suffering from generalized myasthenia gravis in whom the weakness is not irreversible or the patient elderly or ill from other disease. *The ever-increasing dosage of neostigmine and lessened response to it is* an added indication, for it suggests the presence of a thymoma. If a tumour is verified by radiography, operation is undertaken after X-ray therapy.

Mild, localized and easily controlled myasthenia in which ocular symptoms predominate may be kept under observation until such time as evidence of increasing severity or spread manifests itself.

The best results of operation are gained in females under 40 who have had myasthenia for less than 4 years and who respond well to neostigmine. Less good results are gained in others, while at least the progress of the disease is stopped in long standing cases.

Pre-operative Considerations and Care

The myasthenic patient, although co-operative and eager for surgical help, is often anxious, fearful and inclined to panic. It is important therefore that she be admitted into hospital at least a week before the operation is undertaken. During this period of observation, confidence is gained, diagnosis verified and the optimum dosage of neostigmine or mestinon established.

Assessment of Severity of Myasthenia. Soon after admission the patient, with due warning and explanation, is taken off her anticholinesterase drug for approximately 18 hours, although the period would be less if the patient became seriously distressed or helpless. A full examination and test of muscle power as outlined above is then made and

recorded. All muscle groups are tested and if some power remains, retested against resistance and after repetitive exercises.

A cholinergic drug is then injected, either neostigmine 1-2 mgm. intramuscularly or the rapidly acting tensilon, 10 mgm. intravenously. A syringe containing 1/100 of atropine should be in readiness in case of cholinergic gastro-intestinal side effects. The patient is re-examined 10-30 minutes after the injection of neostigmine and the increased scale of muscular activity and any residual weakness recorded. The procedure would prove the presence or otherwise of myasthenia gravis, disclose its severity and would show it to be localized, bulbar or generalized in type. The more complete the response to the injection of neostigmine the higher the scale of activity and the greater can be the confidence in regard to control of the myasthenic state during the post-operative period.



FIG. 227. X-ray showing thymoma.

Residual muscular weakness after an adequate injection of neostigmine shows evidence of some irreversibility of the myasthenia and will be an added anxiety. It is important that the patient regains sufficient power to produce a sharp, explosive cough, otherwise the inability to expel plugs of mucus from the bronchi could lead to post-operative collapse of the lung.

The patient is maintained before the operation on the maximum dose of neostigmine that can be tolerated without side effects. This will bring the muscular strength and scale of activity to as near normal as possible. In an average degree of myasthenic weakness, two or three tablets of either neostigmine or mestinon are given two hourly, the dose to be lessened or increased depending on the response and scale of activity gained.

Electromyography by demonstrating the presence or otherwise of a myopathy or organic state of muscle should be of value in regard to prognosis of the anticipated extent of recovery after thymectomy.

X-ray Examination. It is important that an X-ray of the chest is undertaken before the thymectomy is performed to detect if possible the presence of a thymic tumour. If such a tumour is detected the operation is postponed until after the patient has had a course of deep X-ray therapy. Pre-operative X-ray therapy to the mediastinum generally improves the results of thymectomy when a thymic tumour is associated with myasthenia gravis and does not appear to increase the technical difficulties of the operation.

The normal thymus gland or that of myasthenia gravis cannot be demonstrated on an X-ray film. The majority of thymic tumours associated with myasthenia gravis, however, are relatively opaque to X-rays (Fig. 227) and can be best seen in a lateral view of the anterior mediastinum lying on the aorta or on the pericardium. Postero-anterior views are less helpful but tomography is of value.

Preparation for Operation. Instruction in and practice of deep breathing exercises for several days before the operation.

X-ray and tomographic examination of the chest.

The patient is brought by means of neostigmine to the highest scale of activity possible.

An enema must never be given to a patient suffering from myasthenia gravis. Such an administration may result in a severe collapse or syncopal attack, the patient becoming unconscious, cold, pallid and clammy. This temporary collapse may be due to the stimulation of a parasympathetic system already sensitive and overactivated by cholinergic drugs.

On the Day Previous to Operation. The site for operation is prepared.

A light diet is given.

A glycerine suppository, if necessary, is ordered.

Medinal (barbital sodium) is given at night.

On the Morning of Operation. 2 or 3 tablets of mestinon or neostigmine given 3 or 4 hours before operation.

A further skin preparation is made.

Omnopon 20 mgm. (1/3 grain) and scopolomine 0·4 (1/150 grain) is injected one hour before operation.

Neostigmine 2 ml. (1 mgm.) or mestinon (3 mgm.) is injected half an hour before operation.

A further 2 ml. of neostigmine with atropine sulphate (1/75 grain) is injected 10 minutes before the patient leaves the operation theatre.

It is our experience that a continuous intravenous infusion of saline must not be given during the operation because of the danger of œdema and collapse of the lung.

Preliminaries to Operation. Position of the patient (Fig. 228).

An adjustable bridge which is attached to the operating table is raised under the patient's shoulders, which extends the neck and brings the thorax forward.

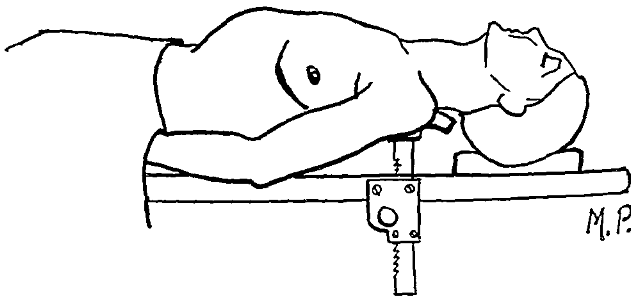


FIG 228. The position for thymectomy.

The required position can also be obtained by means of a sand-bag or cushion placed under the shoulders.

The Anæsthetic. Avoid agents with curariform properties. Induction is by intravenous pentothal and continued by oxygen and cyclopropane. Irritation is reduced to a minimum by avoiding ether and endo-tracheal tubes. The anæsthetic is given through an efficient airway and well fitting mask held in position by a rubber harness. The anæsthetist will inflate the lungs when called upon by the operator who can then easily define and thereby avoid injury to the pleura.

The anæsthesia is light in the final stages from which the patient can quickly recover and rapidly resume a normal, adequate respiratory excursion.

Sub-cutaneous Injection of Adrenaline in Saline. Using a continuous action syringe with a 3-in. needle, the subcutaneous tissues from above the suprasternal notch and over the sternum down to the level of the fifth interspace are injected with a solution of adrenaline in normal saline (1/200,000) in order to minimize bleeding from small vessels. The injection of approximately 100 ml. is given a few minutes before the incision is made.

Thymectomy

The Skin Incision (Fig. 229). A 2-in. horizontal incision is made just above the suprasternal notch extending down to the deep fascia. A further incision commences from the middle of the horizontal incision and extends vertically down the mid-line of the sternum for approximately three-quarters of its length, or to the level of the fifth or sixth rib. The incision is carried down to and divides the periosteum. Skin towels are applied and clipped to the cut margins of the skin.

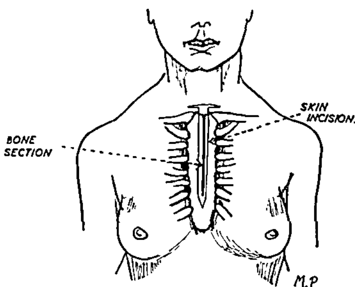


FIG 229 Diagram showing skin incision and line of sternum section in thymectomy

Cervical Access to the Anterior Mediastinum. The deep cervical fascia is opened by a vertical mid-line incision and the sternohyoid and sternothyroid muscles are retracted. This exposes the lower border of the thyroid gland and frequently the cornua of the thymus gland. In this plane the forefinger is forced to its full length directly under the sternum and into the anterior mediastinum, separating any adherent fascia and pushing the pleura laterally so that it will not be injured when the sternum is divided.

The transverse suprasternal ligament is now divided with a knife. Using a Hey's saw the periosteum and the anterior table of the sternum are sawn down the midline for the length of the skin incision (Fig. 229).

Splitting the Sternum. The lower blunt blade of a Sauerbruch's sternum splitter (Fig. 230) is inserted through the cervical opening into the mediastinum, the upper sharp blade being placed in position in the previously incised periosteum. A clean sharp bite

Preparation for Operation. Instruction in and practice of deep breathing exercises for several days before the operation.

X-ray and tomographic examination of the chest.

The patient is brought by means of neostigmine to the highest scale of activity possible.

An enema must never be given to a patient suffering from myasthenia gravis. Such an administration may result in a severe collapse or syncopal attack, the patient becoming unconscious, cold, pallid and clammy. This temporary collapse may be due to the stimulation of a parasympathetic system already sensitive and overactivated by cholinergic drugs.

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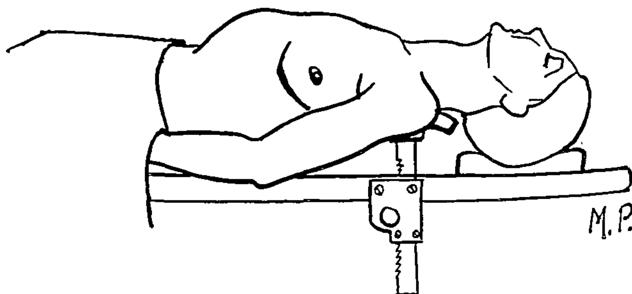


FIG 228. The position for thymectomy

of each lobe is separated from the left innominate vein. Very rarely one of the lobes may lie under it. It is at this stage that care must be taken to identify, ligate and section the main thymic vein ("Great vein of Keynes") which drains into the left innominate vein. Several small arteries are also encountered passing obliquely downward and inward from the direction of the thyroid axis. With these vessels ligated and sectioned the thymus gland can be further turned downwards while a forefinger can safely strip its deep surface

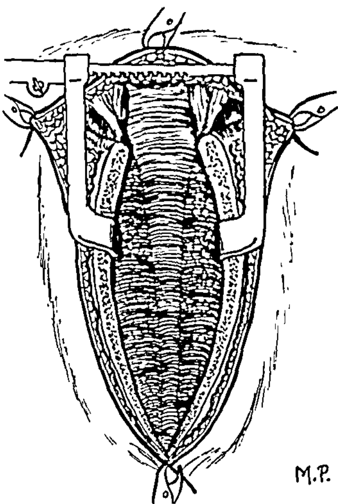


Fig 231. The appearance of the anterior mediastinum after splitting the sternum.

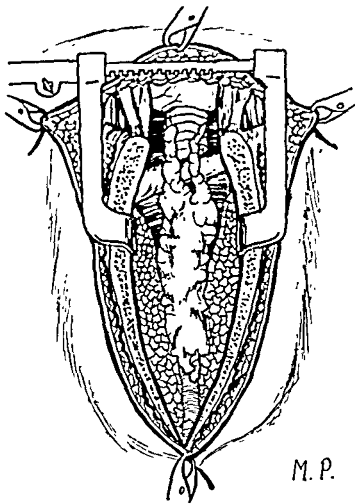


Fig 232. The connective tissue has been incised and the thymus gland exposed (The lungs and pleura lie to either side, the left innominate vein, aorta and pericardium behind)

from off the aorta. The cornua, the lateral borders and the posterior surface of the gland having been freed, all that now remains is to strip the lowest portion of the gland from off the pericardium. As it is most important that the pleura should escape injury, meticulous care must be taken during every stage of the dissection. If by mischance it is opened, a sucking noise would be heard on inspiration and the anæsthetist immediately warned to fully expand the lungs. A small hole would then be ligated or a larger tear sutured with a *running* stitch of fine catgut.

When the thymus gland has been removed, the remaining cavity (Fig. 233) bounded by the aorta, pericardium, lungs and innominate vein is inspected for oozing. Any collection

of the sternum is then made and the instrument is removed. The assistant now forcibly retracts the cut edges of the sternum while the operator explores with his finger to assure himself that the pleura is safe, and then re-inserts the sternum splitter for a further bite. Several bites are taken in this manner, the sternum being gradually divided to the required length (Fig. 229).

Bleeding from the cut surfaces is stopped by pressing small pledglets of Horsley's wax into the cancellous bone. Pleura and connective tissue are next stroked to either side by

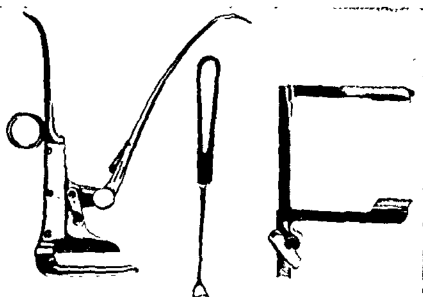


FIG. 230 Sauerbruch's sternum splitter, Joll's thyroid retractor and Tuffier's self-retaining retractor.

swabs of moist gauze and a strong self-retaining retractor (Fig. 230) inserted and adjusted to allow for a good exposure.

Exposure and Dissection of the Thymus Gland. With the divided sternum adequately retracted, the first view of the anterior mediastinum is that of blood-stained connective tissue (Fig. 231). This tissue, which is under tension from the retraction, is cut longitudinally with scissors and as it separates to either side the pleura falls away with it and the anterior aspect of the thymus gland is exposed. The next procedure is to expose fully the anterior surface and lateral margins of the thymus (Fig. 232) and then to dissect it free from the surrounding connective tissue, from the pleura, the aorta and the pericardium, identifying, ligating and sectioning the small blood vessels as they appear.

Much of the exposure and freeing of the gland is best undertaken by blunt dissectors. The smooth pinkish yellow glandular surface of the thymus is picked up by blunt dissecting forceps, while with yet a further pair the connective tissue and the pleura are peeled away with great care and after close scrutiny. When uncertain as to its whereabouts, the anæsthetist is asked to inflate the lungs, after which the pleura and the mottled lung beneath it bulge forward and are quickly identified.

The upper poles or cornua of the gland are now identified and exposed extending up into the neck, often to the level of the lower border of the thyroid isthmus. Each cornu has a blood vessel entering near its apex which is ligated and sectioned to free each pole and to allow them to be turned downwards. As they are turned down, the under surface

The sternal skin incision is closed with nylon, the cervical with horsehair sutures and a few clips. The dressing consists of sterile gauze held in position by a few narrow strips of elastoplast (Fig. 234).

Removal of a Thymic Tumour (Fig. 235) The above description of a thymectomy applies also to the removal of a thymic tumour. If the tumour was diagnosed by means of X-rays, a pre-operative course of deep X-ray therapy would have been given.

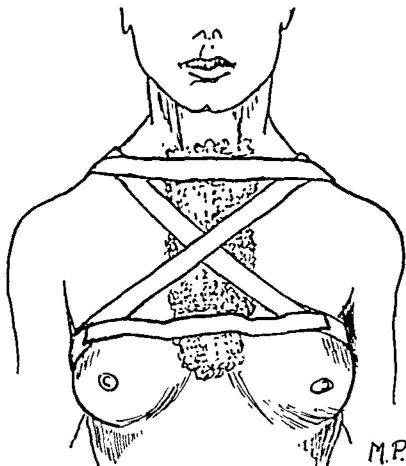


Fig 234 Sterile gauze held in position by a few narrow strips of adhesive strapping

With the divided sternum adequately retracted the thymic tumour is easily identified (Fig 235) as it is firm or hard in consistency, irregular in outline and quite unlike the soft glandular consistency of the rest of the thymus (Fig. 225). It commonly varies in size from a walnut to that of a tangerine orange.

The thymus is [illegible]
the t [illegible]
thickened pleura from off the surface of the tumour before it is delivered. If this is found to be impossible, the lung is fully distended by the anæsthetist and the adherent pleura excised with the tumour. If it is not then possible to close the pleural cavity, the lung is kept distended while the mediastinum is being closed. The removal of a thymic tumour from the aorta or from the pericardium is usually a straightforward procedure, although, on occasions, a portion of the pericardium may have to be excised with the neoplasm. It

of blood is removed with a damp swab, after which the retractor is removed and the cut edges of the sternum allowed to fall together.

Closure of Mediastinum and Fixation of Sternum. Each cut edge of the sternum is bored obliquely down to its inner table; a piece of No. 2 chromic catgut is then threaded through the perforated end of the *awl* and is withdrawn through the bone with it. This is repeated in line on the opposite side and the two free portions of catgut are ready for

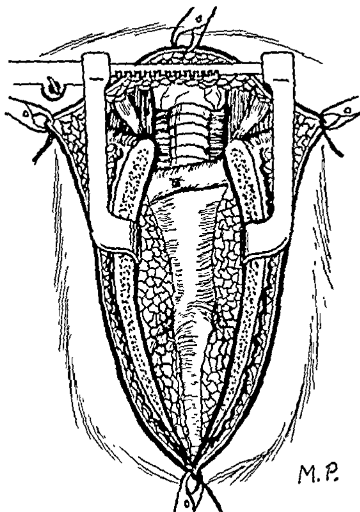


FIG. 233 The mediastinum after the thymus gland has been removed

tying. When this procedure has been repeated at three or four sites, all is ready for the approximation and fixation of the divided sternum. As the cut edges do not automatically fall snugly together, it is necessary for assistants to place closed fists under the patient's shoulders so that the gap will approximate as the bridge is lowered. The chromic catgut sutures are then firmly tied one by one, locking the cut edges closely together. The divided periosteum is next united with interrupted sutures of strong catgut to reinforce the sternal sutures. Before the cervical tissues are finally sutured above the sternal notch, the anæsthetist inflates the lungs in order to force out any blood that may have collected in the mediastinum. This is sucked away by an aspirator. The subcutaneous fatty layer is sutured by fine catgut to lessen the possibility of blood serum collecting.

The sternal skin incision is closed with nylon, the cervical with horsehair sutures and a few clips. The dressing consists of sterile gauze held in position by a few narrow strips of elastoplast (Fig. 234).

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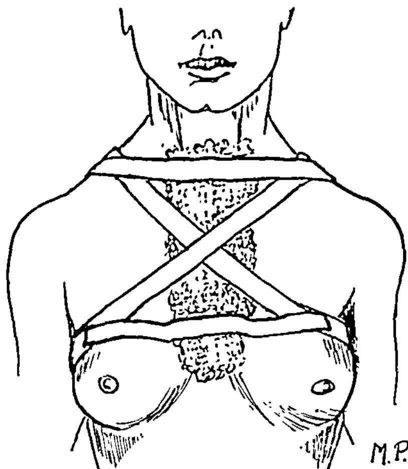


FIG. 234 Sterile gauze held in position by a few narrow strips of adhesive strapping

With the divided sternum adequately retracted the thymic tumour is easily identified (Fig. 235) as it is firm or hard in consistency, irregular in outline and quite unlike the soft glandular consistency of the rest of the thymus (Fig. 225). It commonly varies in size from a walnut to that of a tangerine orange.

The thymus is exposed and freed as described earlier, leaving that portion containing the tumour until the last. An attempt is made, which is usually successful, to peel the thickened pleura from off the surface of the tumour before it is delivered. If this is found to be impossible, the lung is fully distended by the anæsthetist and the adherent pleura excised with the tumour. If it is not then possible to close the pleural cavity, the lung is kept distended while the mediastinum is being closed. The removal of a thymic tumour from the aorta or from the pericardium is usually a straightforward procedure, although, on occasions, a portion of the pericardium may have to be excised with the neoplasm. It

is rare for a tumour to have spread towards the root of the lung or to have invaded the lung. If this has occurred the operation would be abandoned and post-operative deep X-ray therapy relied on.

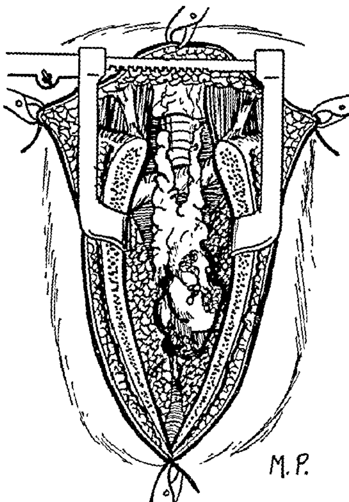


FIG. 235 A tumour of the thymus exposed lying between the lungs and on the aorta

Post-operative Complications

MYASTHENIC OR CHOLINERGIC CRISIS

Myasthenia gravis gives rise to a problem in surgery which is met with in no other condition. It is the danger of post-operative respiratory paralysis, the result of either a myasthenic or of a cholinergic crisis.

Myasthenic Crisis. The weakness of myasthenia gravis is made worse by infection, such as sepsis, dental abscess or influenza. It is also temporarily worsened by the pain and trauma of an operation. The patient with a limited ability to cough because of myasthenia and made still more limited by a mediastinotomy, may be unable to expel a plug of mucus and develop collapse of the lung If by mischance the pleura is opened during the operation, partial collapse of the lung could also occur, creating a desire to cough. A vicious circle is thus set up, leading to an increase in the myasthenic state and possibly culminating in myasthenic respiratory paralysis. To offset this danger, meticulous care must be taken in safeguarding the pleura during the operation and the optimum dose of a cholinergic drug given post-operatively to allow for an explosive cough with

sufficient power to expel mucus. The dose must not be so large as to give rise to side effects, as this would then be overdosage and could lead to cholinergic paralysis.

Cholinergic Crisis. Cholinergic drugs, when given in doses beyond their maximum effectiveness, give rise to cholinergic poisoning and paralysis of the voluntary muscles. The greatest care must be taken therefore, to recognize the early stages of cholinergic poisoning and to be able to distinguish its weakness from that of myasthenia. Gastro-intestinal disturbances are the earliest signs. It is important, therefore, not to hide these signs after operation by masking them with atropine. Other signs are excessive salivation, cold sweats, fasciculations of the voluntary muscles, anxiety, panic and respiratory weakness. The weakness of myasthenia, on the other hand, is not accompanied by any of the above excepting perhaps anxiety or panic. The differential diagnosis is therefore not difficult if atropine has not been given to cloud the issue. If it has been given and an incorrect diagnosis of myasthenic crisis made, a further injection of neostigmine could result in respiratory paralysis and death.

Treatment

Myasthenic Crisis. Injection of the maximum dose of a cholinergic drug. Atropine to be given if gastro-intestinal side effects appear.

Cholinergic Crisis. Stop cholinergic drugs and give intravenous atropine sulphate 1–3 mgm.

Treatment Common to Both Crises:

(1) Morphia to allay anxiety and panic when present.

(2) Oxygen.

(3) If the crisis has become irreversible and does not respond to drugs and the cough remains non-explosive and ineffectual in expelling bronchial secretions, bronchoscopy and suction should be performed.

(4) If respiratory paralysis has set in, with or without pulmonary collapse, immediate tracheotomy must be performed and suction instituted to help maintain a clear airway. Continuous oxygen must be given, for any prolonged cerebral anoxæmia may result in coma. It will be necessary to give manual artificial respiration or provide positive pressure respiratory assistance. The Beaver respirator is an excellent apparatus and is attached to the tracheotomy tube. It provides controlled rhythmic respirations with percentage oxygen for the unconscious or paralysed patient.

At such time as the paralysis comes under control of drugs and consciousness and strength are to some extent regained, the respirator is detached and respiration allowed to proceed normally. The tracheotomy opening is retained for a few days to allow for the suction of bronchial secretions.

Pneumothorax or hæmothorax. These will occur if there is an unrecognized tear of the pleura. Routine portable X-ray of chest 12–24 hours after the operation would confirm.

Treatment for hæmo-pneumothorax, if causing symptoms—aspiration of pleural cavity.

Pulmonary infections. These show themselves about the third or fourth day.

Post-operative shock. This is never severe.

is rare for a tumour to have spread towards the root of the lung or to have invaded the lung. If this has occurred the operation would be abandoned and post-operative deep X-ray therapy relied on.

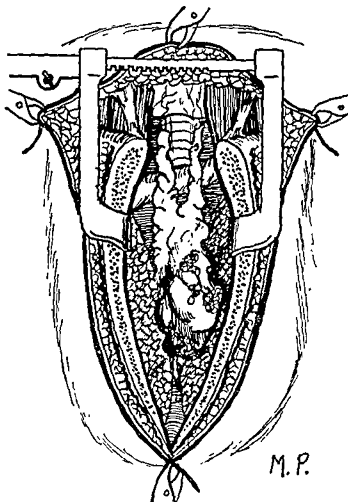


FIG 235 A tumour of the thymus exposed lying between the lungs and on the aorta

Post-operative Complications

MYASTHENIC OR CHOLINERGIC CRISIS

Myasthenia gravis gives rise to a problem in surgery which is met with in no other condition. It is the danger of post-operative respiratory paralysis, the result of either a myasthenic or of a cholinergic crisis.

Myasthenic Crisis. The weakness of myasthenia gravis is made worse by infection, *such as sepsis, dental abscess or influenza. It is also temporarily worsened* by the pain and trauma of an operation. The patient with a limited ability to cough because of myasthenia and made still more limited by a mediastinotomy, may be unable to expel a plug of mucus and develop collapse of the lung If by mischance the pleura is opened during the operation, partial collapse of the lung could also occur, creating a desire to cough. A vicious circle is thus set up, leading to an increase in the myasthenic state and possibly culminating in myasthenic respiratory paralysis. To offset this danger, meticulous care must be taken in safeguarding the pleura during the operation and the optimum dose of a cholinergic drug given post-operatively to allow for an explosive cough with

Portable X-ray of chest 24 hours after operation is routine.

Penicillin is given at the slightest indication of infection.

First post-operative day—sitting up in bed and swallowing tablets by mouth.

Second day, skin stitches and clips removed from neck. Light diet. Mist. A.P.C. or codis for pain.

Fifth day, skin stitches removed from chest. Patient sitting out of bed.

Cholinergic tablets are gradually lessened in accordance with increase of strength.



FIG. 238 Severe myasthenia gravis of 8 years duration in a patient of 50.



FIG 239. The same patient after a large dose of neostigmine showing a poor response due to irreversible changes in the muscles.

The operative mortality in thymectomy is approximately 4 per cent, the direct cause of death being due mainly to pulmonary infections or respiratory paralysis.

It must be appreciated that a patient suffering from myasthenia gravis is particularly liable to pulmonary infection and collapse and that such infection increases the severity of the myasthenia gravis. It follows, therefore, that every precaution must be taken to keep the patient free from infection and that those having contact with the patient must wear masks for several days after the operation.

Careful watch is kept to maintain the maximum level of strength with the optimum dosage of neostigmine and to recognize and treat impending myasthenic or cholinergic crises. An enema must not be given for fear of inducing a cholinergic crisis nor must any form of intravenous infusion be given as a routine for fear of increasing the liability of œdema and collapse of the lungs.

A quick recovery from the anæsthetic is important, as it allows for an early resumption of an adequate respiratory exchange and an early return to oral administration of the cholinergic drugs.

Hæmorrhagic serum. This may collect in the mediastinum, seep through the sectioned sternum and give rise to a fluctuating swelling under the skin. This should be aspirated after the fifth or sixth day.

Post-operative Treatment. The patient is nursed flat until recovered from the anæsthetic, when two pillows are allowed.



FIG. 236 An averagely severe case of myasthenia gravis



FIG 237 The same 10 days after thymectomy.

The dose of neostigmine required after operation by intramuscular injection will depend upon the optimum pre-operative dose that was being taken by mouth. One milligram of neostigmine by injection has the same cholinergic effect as 30 mg. orally. As an example, a patient who required 30 mg. of neostigmine two-hourly before operation would require 1 mg. by injection two-hourly after operation.

On regaining consciousness the patient is encouraged to take deep breaths and to cough. Neostigmine is given by mouth as soon as tablets can be swallowed.

Close watch is kept for evidence of increasing muscular weakness and in particular any lessened ability to produce a sharp or explosive cough. More neostigmine is required if such weakness becomes apparent.

Close watch is kept also for side effects due to overdosage of the cholinergic drug, such as gastro-intestinal stimulation, cold sweats, excessive secretion and respiratory weakness. Neither strong sedatives nor atropine should be given as they will mask the above early symptoms of cholinergic poisoning. Should such symptoms arise, atropine 1/100 grain intramuscularly should be given and there must be a reduction in the dosage of neostigmine. Pain and panic are relieved by omnopon gr. 1/6 by injection and repeated as required. Heavy sedation is to be avoided.

Portable X-ray of chest 24 hours after operation is routine.

Penicillin is given at the slightest indication of infection.

First post-operative day—sitting up in bed and swallowing tablets by mouth.

Second day, skin stitches and clips removed from neck. Light diet. Mist. A.P.C. or codis for pain.

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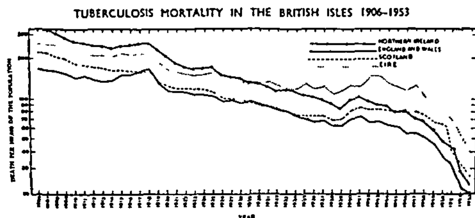
CHAPTER X

THE SURGERY OF PULMONARY TUBERCULOSIS

DILLWYN M. E. THOMAS

INTRODUCTION

THE rapid fall in the mortality rate for pulmonary tuberculosis makes it apparent that a great change has occurred in the disease. This fall, which was steady apart from the war years, has been dramatic since 1947 (Fig. 240), but the decline in the incidence of the disease since 1947 is not in any way so dramatic. This is highly suggestive that the decline in the mortality is almost entirely due to improved methods of treatment.



(*Reproduced by courtesy of "Tubercle."*)

FIG. 240

No one will deny that the fall in the death rate has been in large measure due to the outstanding results achieved with the specific chemotherapeutic agents employed in the treatment of pulmonary tuberculosis. The present favourable trend began with the introduction of streptomycin in 1947, and was accelerated by the subsequent addition of para-aminosalicylic acid (P.A.S.) and isonicotinic acid hydrazide (I.N.A.H.). These drugs have completely changed the climate of hospitals treating pulmonary tuberculosis. No longer do the temperature charts show prolonged hyperpyrexia in young adults suffering from the disease. The symptoms of the acute phase are almost absent. Cough and expectoration are minimal; night sweats, laryngitis and gastro-enteritis are now most uncommon.

Some well qualified authorities believe that chemotherapy is so potent that surgical treatment will no longer be required on the scale practised during the last ten years. Others have expressed the belief that tuberculosis no longer presents a surgical problem. They are confident that together with mortality the incidence of the disease will fall rapidly, and that pulmonary tuberculosis will cease to feature largely in medicine. Others equally well experienced are sceptical, and think that an appreciable percentage of the

Results of Thymectomy. The results of *thymectomy for myasthenia not associated with a thymic tumour* are variable and with a mortality of 4 per cent. The best results are obtained when the patients are young and when the duration of the disease is short. One can expect a complete or near complete remission rate of 70 per cent of female patients under 35 years of age (Figs. 236 and 237). The majority of the remainder are improved but still require a considerable dose of neostigmine or mestinon to increase their scale of activity sufficiently to enable them to work. In 10 per cent of cases there would be no apparent improvement; there is the likelihood, however, that the progress of the disease has been stopped. In this latter category would be those patients of over 45 years of age and with many years' history of the disease (Figs. 238 and 239). Many patients with only a poor or a fair early result find their scale of strength and activity steadily improving as time goes by.

The results of *thymectomy* for myasthenia associated with a thymic tumour are far less good although they have been more encouraging since the practice of giving pre-operative deep X-ray therapy to the mediastinum. Thymectomy nevertheless is strongly indicated in view of the potential malignancy of the tumour.

In some cases of myasthenia gravis the diagnosis is difficult and prognosis as to the degree of improvement or cure is uncertain. The diagnosis and prognosis of such cases in the future should be made far more certain with the aid of recent biochemical and other tests such as decamethonium and electro-myography.

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in daily dosage of 1 g.; 20 g. of para-aminosalicylic acid are given during the twenty-four hours, in doses of 5 g.; 200 to 400 mg. of isonicotinic acid hydrazide are given orally daily, conveniently divided into two or three doses.

Unless there is an urgent need for treatment with chemotherapy, every effort should be made to recover the organism before commencing treatment so that evidence of resistant strains, if present, will prevent the administration of the wrong antibiotics.

The knowledge of the sensitivity of the bacillus is also of value in planning the surgical programme. For example, a thoracoplasty may be chosen in preference to resection when the presence of resistant strains signifies that the antibiotics available will not provide an adequate cover during the operation.

It is current practice to administer antibiotics for six months or even longer prior to resection surgery, and for a further six months after the operation. Antibiotics are not free from toxic effects, a knowledge of which is desirable as they may occur during the post-operative period, giving rise to anxiety and confusion if not recognized.

Toxic Effects

(1) Streptomycin. Dihydrostreptomycin may cause deafness, while streptomycin sulphate is prone to promote vestibular disturbances. Other general reactions such as fever, nausea and vomiting may confuse the post-operative recovery of the patient. Desensitization, starting with small doses such as 10 mg. and working up to 1 g., is often successful.

(2) Para-aminosalicylic Acid. P.A.S. not infrequently gives rise to gastro-intestinal symptoms such as vomiting, nausea and diarrhœa. As with streptomycin, pyrexia and rashes may also complicate the post-operative period. Liver damage with jaundice has been observed and myxœdema reported.

(3) Isonicotinic Acid Hydrazide. I.N.A.H. is not so toxic, but neurological symptoms have been reported.

ADMINISTRATION IN THE IMMEDIATE POST-OPERATIVE PHASE

It has been found a good practical policy to omit wherever possible the administration of P.A.S. during the immediate post-operative period, and use in its stead I.N.A.H. which is less liable to cause vomiting. Many successful resections have been carried out under the cover of viomycin, especially when it can be used in combination with other antibiotics. Viomycin is usually administered in 2 g. doses weekly. The importance of having a potent antibiotic cover during the surgical period cannot be overemphasized, and it is advisable to continue antibiotic treatment for at least six months following a resection.

There are possibly exceptions to the above programme. When surgery is contemplated for tuberculoma, healed tuberculous bronchostenosis or tuberculous empyema, experience has shown that a long pre-operative course of streptomycin will cause unnecessary delay, and that surgery can be carried out after a shorter course of antibiotics.

When antibiotics are administered to infants and small children as a cover for tuberculous surgery, 0·02 g. per pound body weight is the recommended daily dose. I.N.A.H. is better tolerated than P.A.S. in children, and a dose of 100 mg. daily is recommended for children under two, 150 mg. between the age of two and ten, and 200 mg. at the age of ten or over (Roberts, 1954).

cases treated with chemotherapy will relapse in the future. It is apparent, nevertheless, that chemotherapy is altering the surgical approach, and that in most cases a long course of chemotherapy should be administered prior to a decision being made as to the desirability of seeking surgical help.

Rest and Sanatorium Treatment

Rest, so long the sheet anchor of conservative treatment for pulmonary tuberculosis, is still of importance, but there are physicians who suggest that cases of pulmonary tuberculosis can be adequately treated with chemotherapy at home, and non-infectious



(*a*) (*b*)

FIG 241 (*a*), (*b*) These X-rays of a girl aged 19 demonstrate the clearing of acute disease following 9 months' treatment with streptomycin and para-aminosalicylic acid

cases while they are gainfully employed. This, however, is sub judice, and the sputum positive patient is better treated in hospital away from susceptible contacts. Most patients will benefit from rest as they did under the old conservative regime.

Chemotherapy and Pulmonary Tuberculosis

Chemotherapy must be used in adequate doses and in combination for prolonged periods to obtain the optimum therapeutic effect. Streptomycin, para-aminosalicylic acid (P.A.S.) and isonicotinic acid hydrazide (I.N.A.H.) are the most potent and commonly used. Experience has shown that at least two drugs should be used in combination, because so applied they are more effective and also delay the emergence of resistant strains of the bacillus which limit the value of each agent when used alone.

The more acute the disease, the better the chemotherapeutic response (Figs. 241 (*a*) and (*b*)). In chronic disease improvement is not so marked and relapse is common following withdrawal of the drugs. Streptomycin is administered to the adult by an injection

in daily dosage of 1 g.; 20 g. of para-aminosalicylic acid are given during the twenty-four hours, in doses of 5 g.; 200 to 400 mg. of isonicotinic acid hydrazide are given orally daily, conveniently divided into two or three doses.

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It cannot be too strongly emphasized that the administration of antibiotics must be under careful control to avoid the emergence of resistant strains. When this occurs, not only has a disservice been rendered the patient, but infection of susceptible contacts by resistant strains of the bacillus can severely affect the course of the disease in them and jeopardize both conservative and surgical treatment. Haphazard antibiotic therapy is to be deprecated.

Postural Dependency

Another development has been the use of postural dependency in the treatment of cavities (Thomas, 1955). This method helps in promoting cavity closure and minimizes the risk of contralateral spreads, which are liable to occur during sleep (Helm, 1951).

The Principles of Postural Dependency. The writer noted that there was a tendency for a tuberculous cavity to decrease in size when placed in a dependent position. Experience has demonstrated that, when postural dependency is combined with chemotherapy, apparent closure of a tuberculous cavity is achieved in the overwhelming majority of cases.

Methods of Dependency Treatment. The cavity is first localized with lateral X-ray films. Occasionally tomography is necessary for satisfactory localization. The patient is then nursed in a plaster cast, the method varying with the position of the cavity.

(1) CAVITY IN THE APICAL SEGMENT OF THE LOWER LOBE. The foot of the bed is raised on eight-inch blocks, and the cast tilted thirty degrees towards the diseased side.

(2) CAVITY IN THE APICAL SEGMENT OF THE UPPER LOBE. Here, the foot of the bed is raised twelve to eighteen inches, and the cast tilted to about forty-five degrees to the diseased side.

(3) CAVITY IN THE ANTERIOR SEGMENT OF THE UPPER LOBE. Here, the patient is nursed face-down on a plaster cast with the foot of the bed raised four to eight inches, and tilted about twenty to thirty degrees to the diseased side.

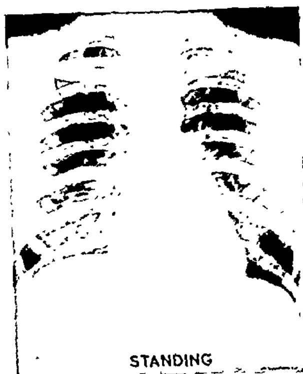
The suggested positions are the approximate optimum for dependency nursing.

The only complication of note that occurs during this treatment is a silent atelectasis, there being neither complaint from the patient nor elevation of temperature or pulse. When this occurs, re-æration of the lung can be rapidly achieved by resuming normal posture.

It is of interest to speculate how postural dependency reduces the size of tuberculous cavities. Radiological studies of healthy people in the recumbent lateral position show that in quiet respiration the dependent diaphragm assumes a position approaching that of expiration, while the uppermost diaphragm assumes the position of inspiration. The dependent lung is small and has the same appearance as in expiration, while the uppermost lung is large, and its size is that seen in inspiration (Fig. 242 (*a*), (*b*) and (*c*)). It is felt that this is a purely mechanical change, and has nothing whatsoever to do with the function of the dependent lung in comparison with that of the uppermost lung. There is some evidence to suggest that during quiet respiration the bronchi in the dependent lung are shortened and dilated, while the bronchi in the uppermost lung are lengthened and narrowed.

The effect of posture on the lung and diaphragm has been explained in detail because the application of this knowledge has been used with benefit in the post-operative treatment of cases of pulmonary resection.

(a)







(b) (c)

FIG 242 X-rays showing the change in the position of the diaphragm. (a) Standing; (b) Lying on the left side, (c) Lying on the right side. Note the difference in size between the dependent and the uppermost lungs.

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(1) Cavity in the Apical Segment of the Lower Lobe. The foot of the bed is raised on eight-inch blocks, and the cast tilted thirty degrees towards the diseased side.

(2) Cavity in the Apical Segment of the Upper Lobe. Here, the foot of the bed is raised twelve to eighteen inches, and the cast tilted to about forty-five degrees to the diseased side.

(3) Cavity in the Anterior Segment of the Upper Lobe. Here, the patient is nursed face-down on a plaster cast with the foot of the bed raised four to eight inches, and tilted about twenty to thirty degrees to the diseased side.

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of stairs will usually suffice to ascertain whether, for example, a pneumonectomy can be undertaken with safety.

(2) Physiological

(*a*) MAXIMUM BREATHING CAPACITY. This will show whether the patient is capable of normal or subnormal ventilation, i.e. whether he has a good pair of bellows. It demonstrates not whether he is capable of normal oxygen absorption, but only the intake of air into his lungs. An adult patient with an M.B.C. of 40 litres per minute is dyspnœic while at rest. This test requires active co-operation to obtain informative results.

(*b*) BRONCHOSPIROMETRY. This measures the function of each lung in terms of ventilation and percentage oxygen uptake occurring in each lung. The following are two examples of the information obtained from use of the above tests.

EXAMPLE I. E.A., aged 48 Fig. 243 (*a*). (*b*) Developed pulmonary tuberculosis twenty years ago and had a left artificial pneumothorax for four years. Difficulty in expanding lung, but patient remained well until three years ago when she developed a left effusion and symptoms of dyspnœa.

January, 1953.	M.B.C. 63 l/min.	
	Rt.	*Lt.*
	%	%
O_2 Uptake . .	90	2
Ventilation . .	96	4

February, 1953: Left decortication, with smooth post-operative course. Now feels very well.

August, 1954.	M.B.C. 94 l/min.	
	Rt.	*Lt.*
	%	%
O_2 Uptake . .	73	27
Ventilation . .	69	31
Vital capacity . .	68	32

After twenty years of pleural thickening, decortication changed the function of the left lung from virtually nil to more than half the normal value.

EXAMPLE II. Fig. 244. This shows an X-ray of a man aged 34 treated for left upper lobe disease in 1942 with a 7-rib thoracoplasty. He remained well until 1955 when he developed a segmental lesion in the right upper lobe. The possibility of surgery was raised, and it was necessary to determine the loss of function due to disease and thoracoplasty in his left lung.

Function:	M.B.C. 138 l/min.	
	Rt.	*Lt.*
	%	%
O_2 Uptake . .	73	27
Ventilation	69	31

i.e. function of left lung was approximately half the normal value. These cases show the value of respiratory function tests in selected cases.

PULMONARY FUNCTION

The improvement in prognosis for a patient suffering from pulmonary tuberculosis has made it necessary to assess with greater accuracy the pulmonary function of the patient submitted to surgery. The five year cure is no longer a basis for judging success. To ensure that the patient will be capable of leading a near normal life requires that any surgical treatment must have as its prime objective the careful preservation of the maximum of pulmonary function compatible with surgical cure of the disease. Thus, diaphragmatic paralysis by phrenic interruption, a minor surgical procedure, can cause an

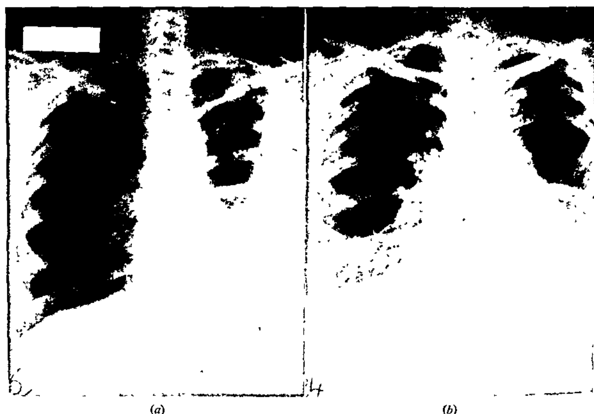


FIG 243 (*a*) X-ray taken before decortication (*b*) X-ray taken 18 months after decortication

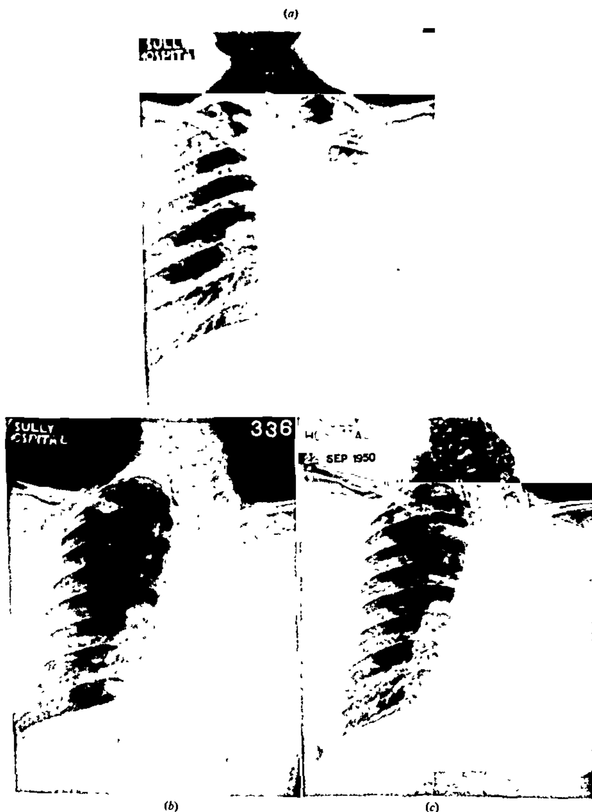
appreciable loss of pulmonary function without compensatory therapeutic results. Conversely, early decortication of a tuberculous empyema may result in a worthwhile improvement of pulmonary function as well as achieving the therapeutic goal of curing the empyema.

Good clinical observation, it is generally agreed, will give an excellent indication of the pulmonary function. Physiological functional tests have, however, a practical value in doubtful cases, and have surgical theoretical importance in demonstrating the degree of functional loss that can be expected from different surgical procedures.

Function Tests in Common Use

(1) **Clinical.** Here the methods employed are simple and consist of observing the respiratory rate at rest and during exercise. The patient's response on mounting a flight

(a)



(b) (c)

FIG. 245. (a) X-ray of ... [illegible]

... well with a negative sputum.

RESECTION FOR PULMONARY TUBERCULOSIS

The acceptance of resection as a form of treatment for pulmonary tuberculosis has been recent. Overholt, Wilson and Gehrig (1952), and others in America; Bickford, Edwards, Esplen, Gifford, Mair and Thomas (1951) and others in this country have demonstrated that resection can be carried out without undue risk, and has a place in the treatment of pulmonary tuberculosis.

Subsequently, in many centres the cases treated by resection far outnumber those who



FIG 244 The function in the left lung underneath thoracoplasty was approximately half the normal value

have had thoracoplasties. This, however, is not universal practice, but represents the general trend.

It is impossible to discuss resection without mentioning the influence of chemotherapy. Now that chemotherapy is more prolonged and its administration produces remarkable clearing of acute disease, its use is limiting the number of cases needing resection.

Indications for Resection

There is no longer any urgency, and no case can be made for resection of florid lesions.

The following indications are generally agreed:

(1) The destroyed lung (Fig. 245 (*a*), (*b*) and (*c*)).
(2) The residual tuberculous cavity. } After chemotherapy.
(3) The residual lesion. }
(4) Tuberculoma (Fig 246).
(5) Tuberculous bronchiectasis.
(6) Broncho-stenosis.

(7) Cavity rupture.

(8) Failed thoracoplasty.

Some of these lesions may be associated with an empyema which is treated concurrently with the intrapulmonary condition.

Pre-operative Treatment. This consists of administration of antibiotics and bed rest and, when a cavity is present, postural dependency helps to reduce its size, so that should a case come to surgery the tendency to a contralateral spread is minimized.

Pre-operative Investigations. The most important investigations consist in determining the presence or absence of resistant bacilli to the common antibiotics, because the best results are obtained when the organisms are still sensitive to the antibiotics.

Next, it is desirable to map out accurately the site and extent of the lesion. To do this tomography, both postero-anterior and lateral cuts, are of value in localizing disease. Pre-operative bronchography with water soluble dionosil may reveal bronchial damage which would influence a surgeon towards a resection rather than a thoracoplasty (Fig. 247).

Bronchoscopy

At one time it was thought advisable to bronchoscope all cases prior to resection. This is now confined to patients considered to have symptoms of active endo-bronchitis or broncho-stenosis of the major bronchi.

NUMBER AND TYPE OF RESECTIONS AT SULLY HOSPITAL IN FIVE YEARS ENDING JUNE, 1953

Operation	1948	1949	1950	1951	1952	1953	Total	Total Mortality
Pneumonectomy	3	22	33	29	23	8	118	5
Lobectomy	—	5	16	23	22	15	81	—
Segmental Resection	—	—	1	2	16	31	50	1
Bilateral Segmental Resection	—	—	—	1	1	2	4	—
Wedge Resection and Spiehectomy	—	—	6	2	2	3	13	1
Total	3	27	56	57	64	59	266	7

Bilateral resections are common, but are usually limited to one or two segments on either side. It must be emphasized that preservation of function consistent with excision of disease is the aim of resection surgery in the treatment of pulmonary tuberculosis.

The Operation

Position of the Table. There is a considerable difference of opinion here. Overholt, Langer, Szypulski and Wilson (1946), Bailey (1947), and Holmes Sellors and Hickey (1949) advocate that the patient should be in the face-down position on the table to prevent contralateral spread. Others, however, consider that the lateral position is perfectly satisfactory, and it has been the writer's experience that, with the use of adequate pre-operative chemotherapy together with postural dependency, contra-lateral spread is uncommon.



FIG 246. Tomograph showing a tuberculoma in the posterior segment of the right upper lobe. This was treated by segmental resection



FIG 247 The bronchogram shows lack of filling of the right upper lobe due to absolute bronchial stenosis, indicating that in this case resection is preferable to thoracoplasty.

custom to use nylon. Whenever possible the bronchus should be covered with a pleural flap. A very good flap can be obtained by stripping the pleura extrapleurally from the apex down to the hilum. The bronchus is covered with this pleural graft, and the mattress sutures threaded through it so that it fits firmly as a cap over the bronchial stump. One of the complications of pneumonectomy, especially a tuberculous pneumonectomy, is the occurrence of an early or late bronchial fistula. The method described above has reduced the occurrence of immediate broncho-pleural fistulæ to a minimum.

Respiratory Function Tests

The maximum breathing capacity is determined in all cases before operation. Bronchospirometry may yield valuable information demonstrating poor ventilation and impaired oxygen uptake on the so-called "good" side. This investigation has an even more important place when the so-called good side has been the site of previous disease treated by means of artificial pneumothorax or diaphragmatic paralysis. Abnormal readings obtained by bronchospirometry can prevent surgery from being carried out on unsuitable cases.

Physiotherapy

Before the operation, the physiotherapist should instruct the patient in diaphragmatic breathing and also in the importance of active post-operative coughing. Upon the appreciation of this and the patient's co-operation depends the smoothness of the post-operative course.

Extent of the Resection

Extent of the resection depends upon the distribution of the disease and the estimated pulmonary function of the residual lung or lungs. When the lung is totally destroyed, pneumonectomy is the only logical procedure. It is of interest, however, that during the last few years there has been a steady decline in the number of cases needing pneumonectomy, and more recently segmental resections outnumber lobectomies. This is well illustrated by Table I (Thompson, Savage, and Rosser, 1954).

The treatment of the divided bronchus after lobectomy or segmental resection is similar, except that one layer of interrupted sutures is sufficient if placed through the uncrushed bronchial wall. Whenever possible a pleural flap should cover the divided bronchus.

When a pneumonectomy has been completed and the bronchus satisfactorily buried by means of a flap, the phrenic nerve is then crushed prior to closure of the chest wall. In contrast, however, remembering the need to preserve function, it is questionable whether the diaphragm should be paralyzed after lobectomy or segmental resection.

Drainage following Resection

Prior to closure of the thoracotomy wound, a drainage tube or tubes are inserted. There is little disagreement concerning drainage following lobectomy or segmental resection, but after a pneumonectomy many think it unnecessary and even dangerous as it may give rise to infection of the pleural space. It is, however, the writer's practice to place a drain into the pleural cavity sited at the posterior axillary line, level with the highest point of the diaphragm, before closure after a pneumonectomy. The tube is removed after 24 hours.

Anæsthesia. The method of anæsthesia varies in different clinics. However, the operation is carried out under general anæsthesia with controlled respiration following the administration of "Flaxedil." When there is evidence of abnormal bronchial secretion, frequent suction during the operation must be maintained.

The Incision. The standard posterolateral thoracotomy approach is used, but should there be any possibility of a secondary thoracoplasty, the incision should approximate the lower two-thirds of the "J," as used in thoracoplasty, thus ensuring that secondary thoracoplasty can be conveniently done by simply extending the vertical arm of the "J." Blood loss is decreased when the skin in the line of the incision, subcutaneous tissues and muscles are infiltrated with a solution of adrenalin, hyaluronidase and normal saline. Usually 400 ml. of this solution is required for a patient of average size. It is prepared as follows: injection of adrenalin B.P. 1/1000 : 1 ml.; Hyaluronidase 1000 Benger units, then made up with normal saline to a quantity of 400 ml. The approach is as for any thoracotomy, the intercostal access being favoured except for patients with a rigid chest. Prior to hilar dissection it is wise to free the lung from any adhesions so that hæmorrhage can be well controlled should the pulmonary artery or any of its branches be inadvertently damaged. This is a wise precaution whether the proposed operation is a pneumonectomy, lobectomy, or segmental resection.

In the operation for pneumonectomy, the arteries, bronchi, and veins are dissected and, if possible, the bronchus is clamped early as this tends to limit contralateral spread. In most lobectomies the arteries are demonstrated and ligated before the bronchus.

In lobectomy when the hilum has been invaded by an inflammatory tissue reaction subsequent to enlargement of tuberculous glands in the primary phase of the disease, the hilar dissection is often hazardous. It is wise then to secure the arteries at segmental level. When divided, the proximal ends of the arteries are carefully dissected away from the bronchus until the desired lobar level of bronchial division is reached. This method of dissection of a fibrosed hilum can significantly reduce the occurrence of severe arterial hæmorrhage. Finally the veins are secured and divided.

In segmental resection the arteries are secured first, then the bronchus. The segment is then teased off the residual lobe in the plane of intersegmental veins. In all resections whenever the bronchus is clamped the anæsthetist should be asked to inflate the residual lung or lungs to ensure that the remaining portions of the bronchial tree are not obstructed by the clamp. During any resection, should a massive hæmorrhage occur from the pulmonary artery, the situation can be recovered by placing a Brock's auricular appendage clamp across the hilum. Control of the hæmorrhage with repair of the artery can then be carried out with greater accuracy and far less loss of blood.

Treatment of the Bronchial Stump

The bronchial stump must be short, although meticulous division at the carina level advocated by many authorities does not appear in practice to be essential. It seems that the stitches should not be placed through crushed bronchial tissue. The actual method of closure is highly individual. After pneumonectomy, the following method has given good results. The bronchus is closed with two layers of sutures; a proximal row consisting of two or three vertical mattress sutures is placed near the carina, and a terminal row of interrupted sutures. Thread, silk, wire and nylon are used for this purpose, and it seems to be a matter of individual choice. For the last four years it has been the writer's

entirely satisfactory, and so the position for post-operative nursing has been considerably modified in the writer's unit.

In young people it has been found that a pneumonectomy patient can be well nursed lying on the operated side. This maintains inflation of the sound side and aids bronchial drainage. It is, however, not well tolerated by patients over 50.

After a lobectomy or segmental resection the patient is nursed on the sound side over a pillow, or better still hyperextended in a plaster cast. This holds the residual lung in a



Fig 249 Showing method of nursing in a plaster cast following lobectomy—to ensure maximum expansion and drainage of the residual lobe and so prevent atelectasis. Note that the side of resection is uppermost

position of inflation and facilitates bronchial drainage (Fig. 249). It is essential, however, that if this routine is carried out continuous suction of approximately half an atmospheric pressure be applied through a water seal via both drainage tubes. Frequent X-rays during the immediate post-operative period are desirable—generally night and morning for the first two or three days, to ensure the early recognition of atelectasis or the development of pleural spaces.

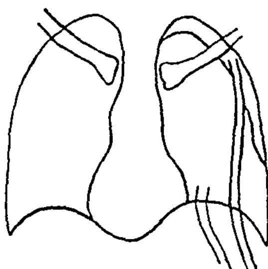
Complications after Resection

(1) **Hæmorrhage.** When bleeding is excessive, clotting is liable to occur, and when gross, re-opening of the chest, evacuation of clot and securing of a bleeding point may be necessary.

(2) **Surgical Emphysema.** This is an indication that the tubes are not effective. Re-adjustment of the tubes may be necessary.

(3) **Atelectasis.** Atelectasis is a direct effect of inadequate productive coughing. If the above modified method of nursing is used atelectasis is uncommon on the operation side, but sometimes there is de-æration of a segment on the contralateral side. When atelectasis occurs, a determined attempt must be made to induce coughing. Inhalation of a cigarette can be effective if smoking is not a habit. The cough is generally inhibited

Rapid re-expansion of the residual lung and pleural space obliteration are imperative after lobectomy or segmental resection. Should this be achieved, the occurrence of fistulæ and empyemata is prevented. As space obliteration depends entirely upon the drainage tubes working efficiently, considerable care must be exercised in their insertion. After much trial and error the following procedure seems the most satisfactory.



(*Reproduced by courtesy of "Thorax," 1954*)

FIG. 248 Diagram showing placing of drainage tubes in the pleural cavity following lobectomy or segmental resection

Two tubes are introduced: one in the posterior axillary line projecting 1 in. into the pleural cavity, level with the highest portion of the diaphragm; another at the same level in the mid-axillary line and passing up to about 2 in. short of the apex. The latter tube must be of good consistency with a thick wall and wide lumen, so that when fixed in position by means of an encircling suture it does not kink. It is also essential that the thickness of the wall of the tube should be such that it does not collapse on the application of high suction. When satisfactorily inserted the tube runs inside but in close apposition to the chest wall (Fig. 248). The opening of the tube is placed at the end, and points upwards so that when suction is applied, the lung is not drawn into the tube, thus destroying the value of suction. When a tube is placed so that the open end is at right angles to the chest wall, the lung is easily sucked into the open end and the value of the suction abolished. After closing the chest, the patient is returned to the ward with the tubes attached to closed under water drainage.

POST-OPERATIVE CARE OF RESECTION CASES

When a tube has been inserted following pneumonectomy resection and attached to an under water drainage bottle, no suction is applied because expulsion of air from the pleural cavity is sometimes sufficient to produce negative pressure so high that there is excessive displacement of the mediastinum with consequent dyspnœa. This undesirable negative pressure must be prevented by using intermittent drainage for short periods only. When an excessive negative pressure has developed in spite of intermittent drainage, dyspnœa and a condition indistinguishable from pulmonary œdema result. When this occurs the momentary introduction of air by disconnecting the tube from under water drainage can be life-saving The tube is removed in pneumonectomy cases after 24 hours.

After lobectomy and segmental resection the drainage tubes should be connected via the drainage bottles with high suction to obliterate the pleural space. The tubes are retained until the pleural space is obliterated and the drainage has ceased together with the expulsion of air.

Position of Nursing. The standard position for post-operative nursing of almost any thoracotomy is sitting up. While this has many advantages, it has not been found

Alveolar leaks are common after segmental resection, and arise from the raw area of the residual lung. They are troublesome and can be minimized by careful oversewing of the raw area on the residual lobe.

When a fistula occurs as a complication to a pneumonectomy, one is faced with a different problem because there is no residual lung to expand and obliterate the pleural space. The symptoms of this occurrence are similar to those described above, and aspiration of any quantity of altered blood into the sound side is serious. On its occurrence, a tube is inserted into the pleural space, the patient nursed on the operated side, and an X-ray taken. If no spread has occurred to the healthy lung, the patient is taken to the theatre, the chest re-opened, and all clot aspirated from the pleural cavity. Whenever possible the fistula is closed and an eight-rib thoracoplasty carried out. A large flap is then fashioned of the thickened parietal pleura, and intercostal bundles and periosteum hinged posteriorly, and placed over the hilar region. The space outside the flap is then drained and the chest closed. The drainage tube is connected to under water suction. The application of this hinged flap of thickened parietal pleura and intercostal bundles, followed by drainage and under water suction, has successfully closed fistulæ even when the bronchi have been so disorganized that closure could not be achieved by sutures.

(6) Empyema. Empyema as a complication to resection is not common when the principle of space obliteration is understood and implemented. When it occurs following lobectomy or segmental resection, drainage, followed by immediate thoracotomy, with decortication and repair of fistula when present, is the treatment of choice. After pneumonectomy, drainage followed by thoracoplasty and a fashioning of flap as described above are usually successful.

The frequency of the complications mentioned above are shown in the following table. The mortality in 266 resection cases is shown in Table II. (Thompson, Savage and Rosser, 1954.)

TABLE II

COMPLICATIONS IN 266 RESECTIONS FOR PULMONARY TUBERCULOSIS (SULLY HOSPITAL FIGURES)

Complications	Pneumonectomy	Other Resections
Major hæmorrhage requiring operation	–	2
Spread	1	6
Leaks	–	15
Broncho-pleural fistula	5	3
Empyema	3	2
Re-activation of disease	4	3

TABLE III

MORTALITY IN 266 RESECTIONS (SULLY HOSPITAL FIGURES)

Operation	Early	Late	Total
Pneumonectomy	3	2	5
Lobectomy	–	–	–
Segmental Resection	1	–	1
Wedge Resection and Spleiectomy	1	–	1

The above results are representative of similar series from many centres.

by pain. The administration of 50 mg. of pethidine given intravenously, followed by encouragement, may produce an effective cough. Should this fail, in the writer's unit a gum elastic catheter is passed through the cords by direct laryngoscopy (Fig. 250). This produces immediate vigorous uncontrolled coughing. A glairy viscid mucus mixed with pus is expectorated with subsequent re-æration of the lung. Bronchoscopic suction can



(*Reproduced by courtesy of "Thorax," 1954*)

Fig. 250. The introduction of a gum elastic catheter into the trachea to produce violent coughing and subsequent re-expansion of atelectatic lung occurring as a complication following either thoracoplasty or resection

produce the same result, especially when the introduction of a bronchoscope gives rise to a spasm of coughing.

(4) **Spread of Disease following Resection.** This is less common in present-day surgery when the cavity is small or closed prior to the operation. When the cavities are large or in the presence of gross tuberculous bronchiectasis, spread of disease is not uncommon. When this occurs, and the bacilli are sensitive to the common antibiotics, clearing takes place following their administration.

(5) Broncho-pleural Fistula, Complicating Resection

With adequate bronchial closure, covering of stump and rapid obliteration of the pleural space, this should not be a common complication. When it occurs, it is made obvious by the coughing of altered blood and the appearance of an air space and fluid level in the pleural cavity. Here it is unwise to temporize. Re-opening of the chest with repair of the fistula either directly or with available pleural flaps, replacing the tubes and the application of suction to obliterate the space have been found to be satisfactory. To temporize by simple tube drainage may result in considerable morbidity.

bronchus was the mechanism by which cavity closure was achieved. This concept of cavity closure has been accepted by many workers. However, Houghton (1950) leads a school of thought which believes that restoration of patency of the bronchus is important, and that bronchial occlusion is not always to the advantage of the patient, especially in cases of artificial pneumothorax.

It is probable that both methods play a part in cavity closure. The results obtained by postural dependency, however, suggest that in cases so treated the open bronchus is an important factor.

Indications for Thoracoplasty

(1) To produce cavity closure when this has not been achieved with chemotherapy and rest.

(2) To maintain cavity closure when it is considered possible that re-opening of the cavity will take place when chemotherapy has been abandoned. Preferably the operation should be carried out in the absence of gross bronchial disease.

(3) Cavities with bronchial disease, where modern combinations of chemotherapy cannot be used because the bacillus is already resistant, to provide an adequate cover for resection.

Ideally, a cavity treated by thoracoplasty should be in the apical or posterior segment of the upper lobe. Cavities in the anterior segment of the upper lobe, or the apical segment of the lower lobe, can be closed only at the expense of considerable destruction of the thoracic cage and pulmonary function.

Secondary Thoracoplasty

Another indication for thoracoplasty is as a secondary operation following resection. A thoracoplasty is used following pulmonary resection to limit the re-expansion of the residual lung, especially where there is some disease in the residual lung; to centralize the trachea so displaced as to cause dyspnœa; and, following a pneumonectomy, to obliterate the pleural space and minimize the formation of late fistulæ and empyemata. The secondary thoracoplasty will be described more fully in the section on thoracoplasty following resection.

Contra-indications

(1) Cavities in the apical segment of the lower lobe.
(2) Other cavities in the lower lobe.
(3) Solid tuberculous foci.
(4) Tuberculous bronchiectasis.
(5) Broncho-stenosis.

Pre-operative Treatment. An adequate course of chemotherapy and rest should precede treatment by thoracoplasty. Should the cavity be large and of the tension type, in the writer's unit it is considered good practice to deflate this cavity prior to operation by nursing the patient on a plaster cast in such a position that the cavity is dependent.

Anæsthesia. The method of anæsthesia varies in different centres. Before the days of controlled respiration and the ready availability of blood, many surgeons turned to local anæsthesia. It is claimed by those who use local anæsthesia that the maintenance of a cough reflex prevents spread to the contralateral side, that the respiration is quiet, and

THORACOPLASTY

The original Sauerbruch thoracoplasty was a sub-periosteal resection of ten ribs in one stage. The mortality was high, and in the main due to paradoxical respiration, which occurs when the bony cage of the chest has been extensively destroyed. It is probable *also that many of the cases were operated upon when the disease was not stable, and the* advantages of blood transfusion and modern anæsthesia were not available. This operation tended to reduce the cavity to a slit, and complete closure of a cavity was only obtained in about 35 per cent of cases.

Later, Holst and Semb, mobilizing the apex of the lung, produced a concentric form of relaxation as opposed to the lateral relaxation of the Sauerbruch thoracoplasty. Concentric relaxation made it possible to achieve a higher rate of cavity closure with less destruction of chest wall. However, it was considered by many authorities necessary to resect sufficient ribs to allow the scapula to fall in. To do this, as many as seven ribs had to be resected, and in more elderly patients sometimes eight. A considerable degree of deformity of the chest wall was a natural sequel to this operation. When the diseased lung has but little fibrosis, or the visceral and parietal pleura is not thickened by the disease process, extensive rib resection and mobilization of the lung give rise to dangerous paradoxical respiration. It therefore becomes necessary to stage the operation, at each stage limiting the amount of rib resection to minimize paradoxical respiration. However, of necessity a multiple stage operation cannot be ideal, for it taxes the fortitude of the patient. Therefore many workers searched for an operation which, while producing the same effect as a multiple stage operation, could be performed in one stage with a minimal production of paradoxical respiration and little deformity.

Thus, many modifications of a thoracoplasty have been presented; Holst, Edwards, Sellors, Brock, and others have all described techniques for a one-stage thoracoplasty.

The Tuberculous Cavity

At this point, before discussing in detail the operation of thoracoplasty, it would be advisable to consider the cause for the persistence of a tuberculous cavity following its formation by the rupture of a tuberculous abscess into the adjoining portion of the bronchial tree.

The bronchus or bronchi draining the cavity become involved in the tuberculous process. The whole mucosa proximal to the cavity may be replaced by tuberculous granulation tissue, but varying degrees of involvement are found, such as sub-mucosal infiltration or sub-mucosal hyperæmia. The lumen of the bronchus so involved is narrowed, and during expiration the stenosis may become almost complete with the result that air which enters the cavity is not entirely expelled during expiration. There is a building up of tension in the cavity which is greater than that of atmospheric pressure. Thus not only is the cavity maintained but it tends to increase in size.

While this is probably not the only mechanism it is considered the most important cause for the persistence of a tuberculous cavity. Relaxation or collapse therapy, whether by artificial pneumothorax or thoracoplasty, is known to cause cavity closure. The mechanism by which this occurs has been the cause of considerable argument. Coryllos and Ornstein (1938) first postulated that complete closure of the draining

bronchus was the mechanism by which cavity closure was achieved. This concept of cavity closure has been accepted by many workers. However, Houghton (1950) leads a school of thought which believes that restoration of patency of the bronchus is important, and that bronchial occlusion is not always to the advantage of the patient, especially in cases of artificial pneumothorax.

It is probable that both methods play a part in cavity closure. The results obtained by postural dependency, however, suggest that in cases so treated the open bronchus is an important factor.

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Anæsthesia. The method of anæsthesia varies in different centres. Before the days of controlled respiration and the ready availability of blood, many surgeons turned to local anæsthesia. It is claimed by those who use local anæsthesia that the maintenance of a cough reflex prevents spread to the contralateral side, that the respiration is quiet, and

that there is a decrease in blood loss. Today, however, it is unusual for a patient to be presented for surgery when there is a large cavity. Pre-operative antibiotics and dependency treatment cause marked reduction in the size of a cavity, and in consequence there is less risk of contralateral spread occurring during the operation, when the cough is abolished under general anæsthesia. The advent of relaxants, controlled respiration and intermittent administration of pethidine has made many surgeons return to the use of general anæsthesia for thoracoplasty. The writer has experience of both techniques, and at present uses general anæsthesia. Should the pleura be inadvertently opened, the use of controlled respiration minimizes all the resultant immediate ill effects. It must, however, be admitted that there is greater blood loss, although limited by infiltration with adrenalin and hyaluronidase along the line of the proposed incision. When blood or a skilled anæsthetist is not readily available, local anæsthesia has a great deal to commend it.

Position on the Table. This is exceedingly important. It is far more important to maintain an exact position for a thoracoplasty than for a thoracotomy. An absolute lateral position must be maintained. This can be achieved by means of chest, pelvic and buttock supports. The position must be so secure that when retraction is applied to the arm and scapula the lateral position is firmly maintained. Any failure to hold this position results in the patient falling prone, which makes resection of the first rib both hazardous and difficult.

Description of the Classical Thoracoplasty with Extrafascial Apicolysis

As the original Semb operation was the precursor of most of the present modifications used, it is described in detail.

The Incision. The incision is planned so that the operation, whether in one, two or three stages, can be completed through the same incision. It is parascapular, and starts at the highest point of the transverse sweep of the trapezius muscle. The vertical part of the incision is continued downwards about 1½ in. lateral to the spinous processes to a point well below the angle of the scapula when it curves towards the mid-axillary line. Should the incision be placed too near the scapula, an ugly scar often ensues; and should it cross the angle, healing is sometimes delayed. Scapula retraction with such an incision is not unduly difficult. The trapezius and rhomboid muscles and the posterior half of the latissimus dorsi are then divided. Next, the upper five digitations and the serratus anterior are freed from the ribs. Then the serratus posterior superior is freed from its insertion into the posterior ends of the fourth and fifth ribs, and finally the scalenus posterior and medius are divided at their insertion into the second and first ribs respectively.

The stage is now set for resection of the ribs. The third rib is resected first, and in a staged operation over a mobile lung it is advisable to remove only the posterior half of the third rib. The rib is divided through the costo-transverse joint posteriorly, and divided anteriorly at about half its length. A similar sub-periosteal resection is then carried out on the second rib. This rib is removed in almost its entire length, the resection stopping short of the second costal cartilage. Sub-periosteal resection of the first rib is then performed, together with division of the scalenus anticus at its insertion, and of the costo clavicular ligament. The first rib is removed from the costo-transverse joint to the cartilage, care being taken not to injure the lower cord of the brachial plexus, subclavian artery or vein.

In order to produce concentric relaxation, it is now necessary to carry out an apicolysis. In the extra-fascial operation designed by Carl Semb, the first stage consists of carefully incising the fibrous envelope of the brachial plexus. The lowest cord is then gently retracted backwards and downwards. Between the plexus and the subclavian artery is found the posterior slip of muscle and tendinous tissue passing from the scalenus muscle in the neck to Sibson's fascia. This is divided. Another such band is found between the artery and the subclavian vein. The subclavian artery is then cleaned, pushing away the pleura and the overlying fascia downwards by blunt dissection. The internal mammary artery arising from the subclavian is followed and also cleaned, at the same time the apex of the lung is mobilized by blunt dissection in the extra fascial plane. Next, the intercostal muscle bundles, vessels and periosteum of the first three ribs are divided posteriorly between ligatures. On the left side the mobilization is carried out until the apex of the lung lies at the level of the arch of the aorta, while on the right side the apex of the lung is mobilized as far as the azygos vein.

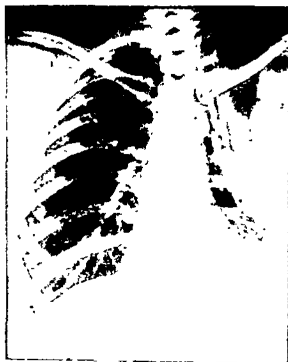


Fig 251 X-ray of a patient with a 7-rib thoracoplasty, which successfully closed a cavity in the apical segment of the left upper lobe.

This constitutes the total sub-periosteal resection of ribs and the extent of mobilization carried out at the first stage. This stage is then completed by closure of the muscles in two layers, with either continuous or interrupted sutures, and then approximation of the skin incision by continuous or interrupted sutures.

The second stage is carried out at the end of 14 days, before regeneration of ribs has occurred. Almost all patients are fit enough to be submitted to a second operation in this short time. The incision is then excised and the muscles opened in the line of their previous division. Most of the remaining anterior half of the third rib is then removed, and decreasing amounts of the fourth, fifth, sixth, and seventh ribs. The extrapleural space produced at the first stage is reopened. The lung is then mobilized posteriorly, again in the extrafascial plane (Price Thomas).

When the lung is very mobile, it is necessary to limit the extent of the second stage by confining resection to the fourth and fifth ribs, together with the anterior end of the third, resecting the sixth and seventh ribs at a subsequent stage.

When the operation has been completed, the scapula should bed itself into its new position with the movements of the arm unrestricted. In older people it is often necessary to remove a portion of the eighth rib to allow this to take place (Fig. 251).

Complications

During the operation by far the most common complication is the accidental opening of the pleural cavity during the mobilization or while resecting ribs. Experience has shown

that the repairing of these tears can be exceedingly tedious and often unrewarding. The insertion of an intercostal tube into the free pleural space in the posterior axillary line level with the uppermost portion of the diaphragm, and the application of suction allow the mobilization to be resumed. Retention of the tube for 24 hours after operation generally results in obliteration of the pleural space.

Injury to the Thoracic Duct. The thoracic duct can be injured during the mobilization. When seen, the duct is secured and ligated. The finding of chylous clots in the extra fascial space during its re-opening at the second operation may be the first indication of duct damage. It is exceptional for the injury of a thoracic duct to give rise to real anxiety.

A Horner's Syndrome. A Horner's syndrome, as shown by ptosis, miosis, enophthalmos, and warm dry hand, due to injury of the sympathetic nerve chain, may be permanent or transient.

Opening of a Cavity During Mobilization. Some lung apices are very difficult to mobilize, the pulmonary fibrosis having extended beyond the confines of the lung. In such cases the cavity may be inadvertently opened. In pre-antibiotic days this was considered a serious hazard. The following procedure, however, was successful in many cases.

The cavity was temporarily closed and the mobilization of the apex completed. When this had been achieved, the cavity was reopened, and closure undertaken by inverting the cavity edges with multiple layers of vertical mattress sutures. Next, using a strip of fascia lying over the lower ribs, an additional covering was made. In some cases where the cavity had been inadvertently opened, the lung was very fibrotic, and the thoracoplasty could be extended by removal of the ribs as far as the seventh to allow the scapula to bed in. This procedure was successful and recovery uncomplicated in three personal cases prior to the days of antibiotics. Should the accident occur now, whenever possible resection of the cavity-bearing area of the lung should be considered preferable to the above procedure.

Hæmorrhage into the Extra Fascial Space. A later complication during the post-operative stage is hæmorrhage into the extra fascial space. A small hæmorrhage is beneficial as it helps to maintain the apicolysis when the lung is mobile. When excessive, however, it can cause pain by tension and later dyspnœa, and may be enough to endanger life. This complication must always be kept in mind and suspected when there is a fall in blood pressure. Tension and dyspnœa can be relieved with aspiration. Should the bleeding continue, however, the space must be explored. In many hundreds of cases the necessity for exploration has been rare The tension, when unrecognized, may be so great as to cause the wound to break down following removal of skin sutures, causing a gaping wound into the extra fascial space. This is most likely to occur 8–10 days after the first stage. When this happens the patient is taken to the theatre, the clot evacuated, and the second stage completed.

Paradoxical Respiration. By paradoxical respiration is meant an indrawing of the chest on the operation side during inspiration and a bulging during expiration. This is a very uneconomical form of respiration, and when severe can be fatal. Severe paradoxical respiration can be partially controlled by firm pads of shaped orthopædic felt strapped over the area of paradox. Extreme paradox, however, denotes that the operation has been badly designed with too much destruction of the chest wall in one stage. Lesser degrees of paradox can be lethal in patients with a poor respiratory reserve.

On regaining consciousness and a normal blood pressure, a patient who has had a thoracoplasty is best nursed sitting up.

Atelectasis. Atelectasis is a danger which must be recognized as a common complication following any chest operation, and efforts must be made to combat it by prevention and rigorous reversal should it occur. Pain, with the suppression of cough, together with paradoxical respiration are the immediate causative agents after thoracoplasty. Cough must be encouraged. Sedatives for the relief of pain are permissible only if a determined effort is made by the nursing staff to make a patient produce an effective cough. While coughing is being undertaken, the operation side should be firmly held by the attendant nurse.

Sometimes it is impossible to overcome a natural desire to avoid pain produced by coughing, and atelectasis then occurs. Coughing must then be forcibly initiated by either bronchoscopy or, as used in the writer's unit, passing a gum elastic catheter between the vocal cords. The latter, when carried out with skill, almost invariably produces a severe bout of coughing and the reversal of atelectasis. Concentration upon the avoidance of atelectasis, and its reversal should it occur, is of paramount importance even in these days of antibiotics. Atelectasis can prove too much for a patient with a poor respiratory reserve, causing an irreversible anoxia.

Persistence of atelectasis is generally complicated by the development of bronchiectasis in the atelectatic lung. When the cause of atelectasis is appreciated and the methods necessary for its reversal understood, persistent atelectasis should be a rare occurrence.

Infection of the Extra Fascial Space. This may be tuberculous or non-tuberculous in origin. When tension develops in the space this should be relieved by aspiration, because rupture of the wound from tension complicates the position considerably. Next, the extra fascial space should be drained by the insertion of a tube into the space from high in the axilla. A fortnight later, with the tube in position, the thoracoplasty should be completed with sufficient resection of ribs to allow the scapula to bed in. The tube is then gradually withdrawn when sinograms show that the space has been reduced to a tube track.

Following a thoracoplasty, a patient is confined to bed for three months. Physiotherapy, however, takes place daily to ensure maintenance of good posture, and freedom of shoulder and scapula movements. The exercises are best carried out when the patient can see fault in posture by looking in a mirror.

The overall mortality for thoracoplasty is approximately 3–5 per cent. It should be remembered, however, that the mortality is increased in patients with a poor respiratory reserve. About 80 per cent of patients achieved cavity closure and a negative sputum. These results were obtained in the days prior to chemotherapy with the standard thoracoplasty.

MODIFIED THORACOPLASTY

The standard thoracoplasty described, although effective in closing the majority of cavities (before the introduction of chemotherapy) has the disadvantage of causing considerable deformity and requiring more than one stage. Further, paradoxical respiration is an ever present danger, especially to the poor risk patient. Any modified thoracoplasty must, while closing cavities with the same efficiency as the standard thoracoplasty, cause minimal deformity, no paradoxical respiration, and require only one operation.

that the repairing of these tears can be exceedingly tedious and often unrewarding. The insertion of an intercostal tube into the free pleural space in the posterior axillary line level with the uppermost portion of the diaphragm, and the application of suction allow the mobilization to be resumed. Retention of the tube for 24 hours after operation generally results in obliteration of the pleural space.

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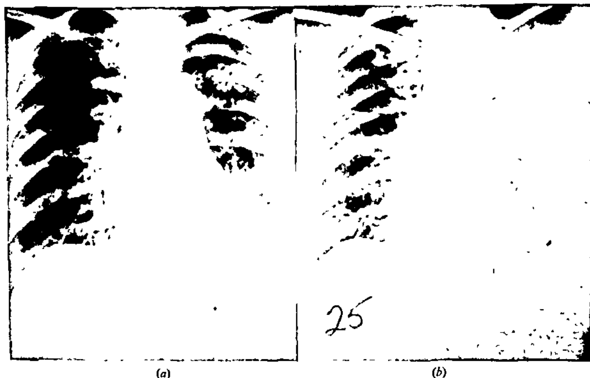
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extrafascial plane. The extraperiosteal space is filled with solid lucite spheres or polythene sponge and the chest closed. Infection of the extraperiosteal space and the occasional necessity for removing the plastic material is a disadvantage in this type of operation (Fig. 252).

A Modified Thoracoplasty Similar to that Described by Holst

This operation is designed to maintain the apex of the lung in its new position by sutures until a new firm osteoplastic barrier has developed to prevent the apex from re-expanding. The approach is similar to that used for a first-stage standard thoracoplasty.



(*a*) (*b*)

FIG 254 (*a*) X-ray showing residual disease in the left lung, especially the upper lobe, and scattered disease in the right lung Considered unsuitable for resection because of the extent of the scattered disease in the right lung The major disease in the left lung contained a small residual cavity. (*b*) Following a Holst thoracoplasty.

Next, about an inch of the posterior ends of the fourth, third, and second ribs is resected periosteally, followed by the dividing of these ribs anteriorly. The periosteum is then stripped from the underneath surface of the first rib and the intercostal bundles divided. When this has been completed the apicolysis is carried out in the normal manner, but the free apex, unlike that in the standard thoracoplasty, has considerable portions of the second, third, and fourth ribs attached to it. Nylon sutures are passed around the posterior ends of the freed second and third ribs, and both ends of each suture passed from within the thorax through the fifth intercostal space as far back as possible. The ends of the nylon sutures are then tied, thus fixing the apex of the lung in its new position (Fig. 253). The chest is then closed (Fig 254 (*a*), (*b*)). Holst divided the second, third, and fourth ribs into a number of pieces and advised the removal of the first rib. This operation goes a long way towards achieving the aim of the modified thoracoplasty, i.e.

Extrapleural Pneumothorax

Extrapleural pneumothorax, while strictly not a thoracoplasty, can be described here. This was first introduced by Tuffier as long ago as 1895. An apicolysis is carried out in the extrapleural plane approached by means of an intercostal incision or by means of the resection of the posterior half of the fourth rib. While having the advantage of a one-stage operation with no deformity or paradoxical respiration, it has serious disadvantages. The collapse is maintained by air refills introduced at positive pressure. The complications are similar to those inherent in artificial pneumothorax, i.e. effusion, cavity rupture,



Fig. 252. X-ray of a patient in whom a bronchial extraperiosteal fistula developed 7½ years after the operation of extraperiosteal plombage with insertion of lucite balls. Note the air gap in the extraperiosteal space.

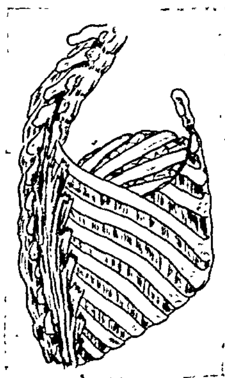


Fig. 253. Diagram of a modified Holst thoracoplasty showing anchoring of the apex in the sixth intercostal space by means of sutures.

bronchopleural fistula and empyema. Attempts were made to maintain the collapse with oil and paraffin wax. More recently the use of inert plastics as a means of plombage led to the development of the extraperiosteal operation (Lucas and Cleland, 1948).

Extraperiosteal Plombage

In this operation the aim is to use the periosteum and intercostal muscles as a barrier between the plomb of lucite spheres introduced into the extrafascial space. Other surgeons have favoured the use of shaped polythene sponge as a plomb.

The approach is similar to that described for thoracoplasty. The posterior ends of the second and third ribs are divided following stripping of the periosteum. Next, the periosteum is stripped off the fourth and fifth ribs and the under surface of the first rib. The intercostal bundles are divided posteriorly and the apicolysis completed in the

THORACOPLASTY AFTER RESECTION

In many instances after resection has been completed, it is considered advisable to carry out a secondary thoracoplasty, often called a corrective thoracoplasty. This secondary operation may be carried out immediately after the resection or at an interval of a fortnight or 3 weeks.

The purpose of a thoracoplasty following a resection is:

(*a*) to reduce space;

(*b*) to prevent over-distension of the residual lung;

(*c*) in the case of a pneumonectomy, to obliterate the pleural space and prevent the formation of a late fistula and excessive deviation of the trachea.

In general, it is considered that over-distension of a lung is detrimental when there is disease in the residual lung or lobe.

Thoracoplasty after Pneumonectomy

It must be assumed that at the site of resection of the main bronchus at pneumonectomy there is, although not apparent, some tuberculous infiltration in the mucosa or sub-mucosal tissues. The pleural space now being empty, the scene is set for the development of a bronchopleural fistula. This may occur between the eighth and the twenty-first day, or months afterwards. Thus, following a pneumonectomy, a secondary thoracoplasty is performed to obliterate this space. Further, there is a tendency for the trachea to be displaced towards the pneumonectomy side. This displacement can be so severe as to cause dyspnœa.

A thoracoplasty not only obliterates the space, but maintains the trachea in a central position. It is said by many authorities that a thoracoplasty also prevents over-distension of the contralateral lung, and that this over-distension diminishes the function of the lung. There is no evidence at present to suggest that the over-distension in point of fact does diminish the function of the lung. However, over-distension is dangerous should there be any tuberculosis in the residual lung. Therefore, the main function of a thoracoplasty following pneumonectomy is to prevent over-distension with subsequent activation of tuberculosis in that lung.

The extent of the thoracoplasty should be sufficient to maintain a central trachea, prevent over-distension, and obliterate the pleural space. While the trachea can be kept central by the removal of the first four or five ribs, it may be necessary to take portions of as many as seven or eight to obliterate completely the pleural space.

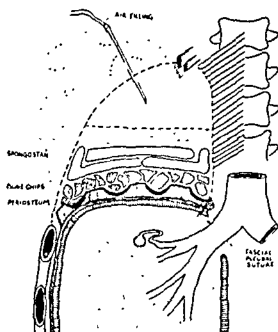
When carrying out this thoracoplasty, some surgeons do not resect the first rib, while others do so although the deformity is greater, as a more certain method of maintaining a central trachea and obliterating the space. It is the writer's custom when carrying out this type of thoracoplasty to remove the first rib, re-open the pleural space, inspect the hilum, and use the thickened parietal pleura and intercostal muscles as a flap over the hilum as an additional safeguard against the development of a late bronchopleural fistula. This extensive operation is carried out a fortnight after the initial resection.

Thoracoplasty following Upper Lobectomy

Should there be no disease palpable in the middle or lower lobes, especially in the apical segment of the lower lobe following a right upper lobectomy, experience has shown

minimal chest deformity, a one-stage operation, and the elimination of paradoxical respiration.

Semb himself has now modified his thoracoplasty so that the rib resection is reduced. In his present operation, having achieved an extrafascial apicolysis, he maintains the collapse and prevents re-expansion using free bone chips from the resected ribs placed over the apex and covered by pads of spongostan. The collapse is further maintained by air refills until a bony roof is formed, solid and resistant, preventing re-expansion of the apex (Fig. 255).



(Diagram reproduced by courtesy of the University of Oslo of the Johan Holst Memorial Volume (1955))

FIG. 255 This diagram shows Semb's method of maintaining the apicolysis with bone chips, spongostan, and temporary refills until the new bony apex becomes quite firm

Other modified thoracoplasties using an osteoplastic flap to maintain the collapse while limiting the amount of rib resection have been described by Sellors, Jackson and Callanan (1955), and Russell Brock (1955). Brock states that the principle of the operation is to perform the correct amount of extrapleural or extrafascial pneumolysis, and then hold the lung in the correct position by an osteoplastic flap composed of segments of the third and fourth ribs, with the intercostal muscles, vessels and nerves intact in front and behind so that the nutrition of the flap is unimpaired. The cut ends of the ribs in the flap are sewn behind to the anterior longitudinal ligament of the spine and in front to the rib cartilages.

At the present time one of the greatest indications for a modified thoracoplasty appears to be as a secondary procedure following resection to reduce space.

Thoracoplasty or Resection?

It is obvious that the indications for resection and thoracoplasty are not clearly defined for lesions in the apical and posterior segments of the upper lobes without bronchial stenosis. Whereas resection would be the choice in some centres, other surgeons prefer to rely on a thoracoplasty. When there is also disease in the contralateral lung or the apical segment of the ipsilateral lower lobe, an even stronger case can be made for thoracoplasty

Resection can only be justified if the incidence of morbidity due to bronchopleural fistula and empyema is small. Resection requires great post-operative vigilance if these complications are to be kept at a reasonable level Nevertheless, resection is the writer's choice for lesions confined to one or two segments, when the operation can be covered with potent chemotherapy, thoracoplasty being reserved for patients with more widespread disease, especially when the contralateral lung is affected.

recommended as a definitive method in the treatment of tuberculous empyema. Temporary drainage in the presence of a bronchopleural fistula is, however, necessary to prevent the flooding of the contralateral lung.

Let us consider the surgical treatment of the case uncomplicated by bronchopleural fistula. Firstly, the condition of the underlying lung must be ascertained; bronchography, bronchoscopy and tomography are of value. Should there be obvious intrapulmonary disease, bronchostenosis or bronchiectasis, excision of the affected area must of necessity modify the operation. The empyema is then drained by means of an intercostal catheter, under local anæsthesia, with the patient in the sitting-up position. The site for this drainage should be well away from the thoracotomy incision. When the empyema has been allowed to drain slowly and no untoward effect has occurred, the patient can then be anæsthetized and placed in the lateral position for a thoracotomy. The skin incision should be of the parascapular type, similar to that made for a thoracoplasty, the vertical part of the incision, however, need not extend so high. This incision allows for the possibility of a subsequent thoracoplasty.

The site of rib resection is generally the fifth, but this may depend upon the extent and position of the empyema. The sub-periosteal resection is often difficult because the ribs are close together and triangular in shape. It is usual to find the parietal pleura thicker than the visceral pleura. While it is tempting to try and excise the empyema intact, it is safer to open the empyema and suck out any remaining pus to prevent contralateral spread caused by an unrecognized bronchopleural fistula.

The opened empyema is next excised. In chronic cases this can be very difficult indeed, and a higher or lower supplementary intercostal incision may be necessary to decorticate the lung, chest wall and diaphragm. A serious attempt should be made to decorticate the diaphragm because a rigid diaphragm means a considerable loss of function.

Stripping the visceral pleura may be exceedingly difficult, especially in the presence of underlying fibrous disease. Multiple alveolar air leaks are common at the end of the operation. When necessary, a lobe, segment or a lung can be excised at the same operation. Though the pleural excision can be extremely difficult, when it has been achieved the hilum in contrast allows of easy dissection. The thorax is then closed and drained, as described following thoracotomy with lung resection (Fig. 256 (*a*), (*b*) and (*c*)).

Post-operative Course

As multiple air leaks are frequently present at the end of the operation, in order to obliterate the pleural space it is necessary to apply high negative suction. The tubes are retained until there is no further drainage of fluid or expulsion of air. The method of post-operative nursing is similar to that described for lobar and segmental resection.

Treatment of a tuberculous empyema by decortication has a double purpose, firstly, the cure of the empyema, and secondly, the possibility of improving the function of the lung when the constrictive pleuritis has been eliminated. Savage and Fleming (1955) report the study of 43 tuberculous patients in whom decortication of the lung was carried out. There was no mortality, and morbidity was minimal. Pre- and post-operative bronchospirometry was performed in 18 cases, and they report that in the majority of these lung function was considerably improved. They advise delaying the test for a year, because while ventilation improves quickly, the oxygen uptake improves more slowly.

that it is not necessary to carry out a secondary thoracoplasty where there is complete re-expansion. The left upper lobe, however, occupies a very large space, and it is considered advisable to perform a four or five rib thoracoplasty to diminish the space left after the removal of this large lobe.

When there is residual disease following a right upper lobectomy and especially when the apical segment of the lower lobe is under suspicion, it is deemed advisable to carry out a four or five rib thoracoplasty to diminish the pleural space and prevent over-distension of the remaining lobes, and so minimize the reactivation of any residual disease. It cannot be emphasized too forcibly that the danger point is the apical segment of the lower lobe.

A secondary thoracoplasty using an osteoplastic flap, as described by Sellors and Brock, has an important application after upper lobectomy, and is an excellent method of reducing space.

In segmental resection such a small amount of functioning lung is removed that a secondary thoracoplasty is generally not necessary. However, this must be modified should the segment removed be the cavity-bearing area only, and much residual disease is left behind. When the pleura is inadvertently opened during a secondary thoracoplasty, the pleural space is drained as low as possible for 48 hours. The complications following a secondary thoracoplasty are similar to those following a primary thoracoplasty.

TUBERCULOUS EMPYEMA

A tuberculous empyema arises as a complication of primary tuberculosis, or following rupture of a tuberculous cavity, or as a complication to treatment of pulmonary tuberculosis with artificial pneumothorax or resection.

In the writer's experience the majority of cases occurred during treatment with artificial pneumothorax. With the decline in the use of artificial pneumothorax the incidence of empyema has fallen.

A tuberculous empyema can cause serious morbidity, especially when complicated by the development of a bronchial or cutaneous fistula. The complicated tuberculous empyema, untreated, inevitably results in death following spread to the contralateral lung or the occurrence of amyloid disease.

Treatment

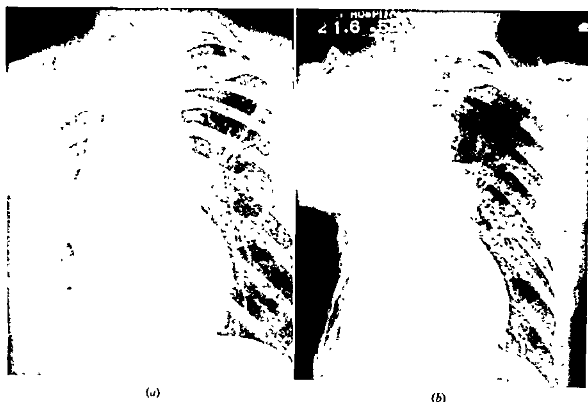
A few cases were successfully treated with aspiration, but unfortunately the majority require more rigorous treatment. A deceptive feature is that the patients remain in comparatively good health until the onset of the complications mentioned. It is pleasing to record that with the decline in the incidence of tuberculous empyema and the improved treatment, amyloid disease is no longer common in hospitals treating advanced cases of pulmonary tuberculosis.

Surgical Treatment

Drainage alone is not often successful because the visceral pleura encases the lung in a cowl of fibrous tissue, which prevents the expansion of the lung and perpetuates the tuberculous empyema. Following drainage the pleural cavity soon becomes secondarily infected, hastening the onset of amyloid disease. Therefore simple drainage is not to be

The results quoted above demonstrate that when adequately treated the prognosis for a tuberculous empyema has changed in a remarkable measure for the better. The operation is carried out under a pre-operative and post-operative umbrella of antibiotics.

When incomplete expansion of a lung, such as persistence of an apical space, takes place following a decortication, it is advisable to obliterate this space by means of a thoracoplasty, especially in the presence of unresected disease of the lung.



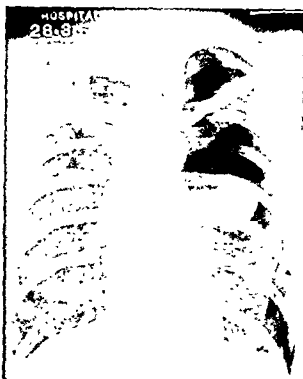
(a) (b)

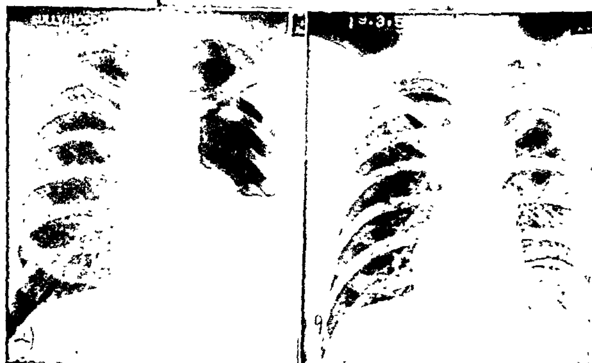
Fig 257 (a) X-ray of a neglected empyema treated for over a year by aspiration and subsequently intercostal drainage when complicated by bronchopleural and cutaneous fistulæ. (b) X-ray when the empyema had been cured by thoracoplasty with obliteration of empyema space by a flap composed of thickened parietal pleura, intercostal bundles and periosteum. Note the deformity and the complete functional loss of one lung.

Prior to the success achieved by decortication in the treatment of empyema, some success was obtained by extensive thoracoplasty. This, however, did not, owing to the rigid nature of the walls of the empyema, always succeed. It was then necessary to carry out a revision thoracoplasty, resecting the regenerated ribs over the empyema. When this was accomplished, the empyema was incised through the thickened parietal pleura. A flap was fashioned of thickened parietal pleura, intercostal muscles and periosteum, hinged posteriorly, and allowed to fall into the empyema space. This operation achieved a great measure of success in curing a tuberculous empyema (Fig. 257 (a), (b)). However, it has the disadvantage that the cure of the empyema resulted in considerable deformity, and at the best no improvement in respiratory function was achieved.

It follows that whenever possible decortication with excision of the pleura is the operation of choice.

(a)



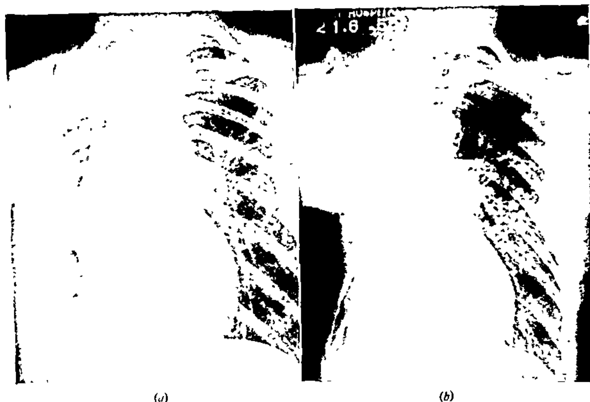


(b) (c)

FIG 256. (a) X-ray showing left upper lobe disease treated by means of an artificial pneumothorax. (b) The artificial pneumothorax is now complicated by an empyema. (c) Following decortication and excision of the apico-posterior segments, left upper lobe In this case the phrenic nerve was also paralysed, but should not be considered a desirable routine measure.

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(a) (b)

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THE SURGERY OF TUBERCULOUS MEDIASTINAL GLANDS

It is generally agreed that primary tuberculosis in children does not give rise to any anxiety in the majority of cases. However, in some cases gross enlargement of the glands of the hilum draining the parenchymal lesion can cause complications due to pressure and erosion of the adjoining bronchial tree. When the glands are enlarged they may progress to caseation and pus formation. When this occurs there is a great tendency for the glandular contents to be evacuated into the bronchial tree. On rare occasions a large paratracheal glandular mass will compress or erode the trachea causing dyspnœa and stridor. Death has been reported from this cause.

A similar erosion can occur into the right or left bronchial tree, and once again the obstruction can be either complete or partial. When complete there is collapse of the lung beyond. Even in partial obstruction the lumen can be so narrowed as to cause complete obstruction during expiration. When this occurs the lung beyond the obstruction becomes over-distended and the mediastinum displaced to the contralateral side with depression of the diaphragm.

In expiratory obstruction, clinical examination will reveal a stridor on expiration, prominent hemithorax, hyper-resonance to percussion, and absent breath sounds. Both Métras (1953) and Thomas (1952) have succeeded, when the obstruction to the trachea and the main bronchial tree by the eroding tuberculous glands has not been relieved by bronchoscopy, following thoracotomy in incising the offending gland mass, evacuating the caseous and purulent material, and thereby relieving the obstruction. When evacuation of the gland mass has been completed, should the gland have actually eroded the tracheal or bronchial mucosa, a fistula communicates with the gland capsule. This fistula can be closed by using the redundant gland capsule as a flap for this purpose (Fig. 258 (*a*), (*b*), (*c*) and (*d*).

In a personal series where over a hundred children with caseous mediastinal glands have been explored by thoracotomy, two children have died, but the morbidity has been negligible in spite of extravasation of tuberculous pus during the operation.

THE PRESENT PLACE OF MINOR COLLAPSE THERAPY IN THE TREATMENT OF PULMONARY TUBERCULOSIS

Artificial pneumothorax was for a long time the most common form of collapse therapy used to achieve cavity closure, but in recent years it has fallen into disrepute and is no longer extensively practised. The induction of an artificial pneumothorax produces a pleural space over the tuberculous lesion, and complications such as tuberculous effusion and frank pus occur in an appreciable percentage of cases. It has also become evident that during the treatment an unpredictable loss of function occurs in the lung because of the gradual thickening of the visceral pleura. The control of a small cavity is achieved often at the expense of considerable loss of pulmonary function. Many of the pneumothoraces are ineffective because the cavity is prevented from closing by adhesions between the visceral and parietal pleura. It is necessary to divide these adhesions to allow the lung to collapse, and so achieve cavity closure.

The operation is carried out by inserting a trocar and cannula intercostally in the mid-axillary line, and another in the posterior scapular line at about the angle of the scapula.

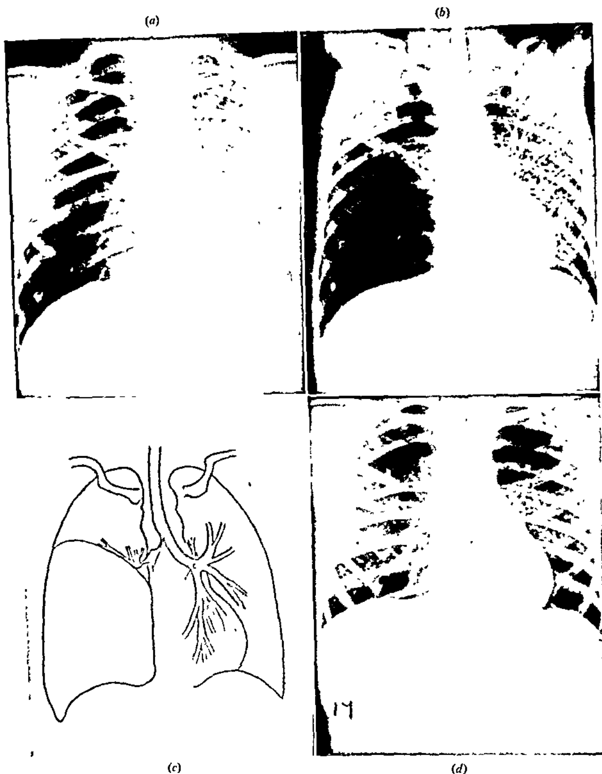


FIG 258 (a) X-ray showing over-distension of the right lung due to obstructive emphysema, causing mediastinal displacement and depression of the diaphragm. (b) Bronchogram demonstrating

is no longer distended

The adhesions are then divided by means of a cautery. The division is observed with a telescope inserted through the second cannula. It is necessary to divide the adhesions close to the parietal pleura, occasionally enucleating them out of the chest wall so as to avoid injury to the lung. To divide short adhesions is considered dangerous. This operation needs a great deal of practice, but has proved of great value in making the pneumothorax more effective in closing the cavity. In adults the operation is carried out under local anæsthesia except in the presence of bilateral pneumothoraces in patients with poor function. In such cases general anæsthesia with controlled respiration skilfully employed prevents such patients becoming anoxic.

Foster-Carter *et al.* (1952) reviewed 457 patients treated by artificial pneumothorax at the Brompton Hospital and its Sanatorium 5 years after their artificial pneumothoraces had been abandoned. They concluded that the "key" to successful pneumothorax treatment was closure of cavities and not the presence of indivisible adhesions. The survival rate in the patients with a free lung was 91 per cent and in the adherent group, but with cavity closure, which represented 242 patients, a survival rate of 94 per cent. These results suggest that the presence of adhesions does not of necessity prevent cavity closure in all cases.

Complications

During the operation severe hæmorrhage occasionally occurs when adhesions are divided, but this is not as frequent as one would suppose.

Post-operative Complications. Coughing and vomiting after the operation tend to produce surgical emphysema which in itself is not harmful, but its occurrence means that the pneumothorax is in danger of being lost because the air has escaped from the pleural cavity into the subcutaneous tissues.

Post-operative hæmorrhage can occur and is generally due to bleeding from intercostal vessels damaged during the insertion of the cannulæ. This, however, does not become obvious until the cannulæ have been removed. When the hæmorrhage is severe thoracotomy is necessary to control it and to evacuate the clot.

Late complications include tuberculous effusion, empyema and cavity rupture. Finally, when the pneumothorax is abandoned, the cavity may re-open or the lung may be incapable of re-expansion.

Operations on the Phrenic Nerve for Pulmonary Tuberculosis

Paralysis of the diaphragm was advocated as a means of cavity closure. This operation is simple to carry out and is an attractive surgical exercise.

The incision is made two fingers' breadth above the clavicle; the posterior border of the incision starts at the external jugular vein and is carried forward for about an inch. When platysma is retracted and the pad of the scalenus anticus muscle is exposed, the nerve is found running across the latter from without inwards. The nerve, still lying beneath the fascia, is then crushed. To ensure that the diaphragm is paralysed, a search is then made for a possible accessory phrenic nerve. The nerve is crushed gently in the hope that the paralysis of the diaphragm will be temporary and not permanent. The practice of phrenic nerve avulsion has been abandoned for many years. Unfortunately, there is no guarantee that the diaphragm will recover its function even after the most gentle crushing.

The advantage of this procedure is that it is not a major operation; serious complications are uncommon, but damage to the brachial plexus, jugular vein, thoracic duct and accessory nerve has been reported.

Disadvantages of the operation are that *in only a small percentage of cases does it* achieve its objective—cavity closure; the paralysis may be permanent and bronchospirometry *shows that the loss of lung function* by this unpredictable operation is appreciable.

It should be pointed out that this loss of function may in some extensive cases of tuberculosis prevent the employment of a more efficient method. In conclusion it is probable that there is little indication for the use of phrenic interruption as a method in the treatment of pulmonary tuberculosis.

CONCLUSION

Ten years ago cavity closure by collapse methods was the most common indication for surgery in the treatment of pulmonary tuberculosis, and a high measure of cavity closure and sputum conversion was obtained, but bronchial disease was largely unaffected by these procedures, and was responsible for many of the failures.

When chemotherapy proved to be a potent cover for resection of tuberculous disease, preventing the lethal complications which had hitherto occurred when tuberculous disease was traversed, lung resection became a practical method of dealing with the bronchial lesion as well as the parenchymal disease.

Today, with the improved results obtained with longer periods of chemotherapy, surgery is directed towards the treatment of the residual disease by collapse methods, or more commonly, resection, but the position is far from clear because in resected specimens tubercle bacilli are almost invariably seen and grown when less than 4 months' preoperative chemotherapy is given. However, after 9 months of chemotherapy, although acid fast bacilli are often seen, they are seldom grown.

The difficulty of interpreting these findings prevents true evaluation of the place of surgery in the treatment of pulmonary tuberculosis. The future behaviour of the residual lesions following withdrawal of chemotherapy will have to be assessed on clinical, radiological and bacteriological grounds. These latter findings will determine the role of surgical treatment in pulmonary tuberculosis.

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CHAPTER XI

THE SURGERY OF THE HEART AND THE GREAT VESSELS

N. R. BARRETT and JOHN ANDERSON

GENERAL PRINCIPLES

THE heart is the last organ to come within the ambit of surgery and the ideas in vogue today date from the end of the Second World War. The reasons why cardiac surgery was not developed sooner do not concern us: but the fact that from 1946 onwards more progress has been made than in all previous time means that ideas are fluid, experiments and theories are dominant, late results are not known, and that no part of this chapter will stand unchallenged.

Cardiac surgery today is in an interim period: it stands between the crude manipulations which were in practice in the past and the accurate surgery of the future. It might be compared with prostatic surgery at the time when Peter Freyer remarked that his "eye was on the end of his finger."

Strange beliefs have prevented surgeons from operating upon the heart in the past; one of these was the idea that it was a delicate structure which could not tolerate surgical intervention. But in fact it is a strong organ. It can function adequately in spite of congenital deformities and acquired diseases; it does not resent surgical incisions or prolonged manipulations; it heals soundly by scar tissue, and, throughout life its various parts, such as the valves, are constantly replaced so that they do not wear out.

Whether the cardiac lesion which requires correction is congenital or acquired the ideal is to deal with it so that an entirely normal state of the valves and great vessels is created. This is clearly a counsel of perfection because many of the conditions which are amenable to surgical treatment are themselves complicated by coronary artery or myocardial disease. The principle which we wish to underline is this: if a surgeon has a choice of two alternatives one of which offers alleviation by adding an extra shunt or a by-pass to the existing anomalies, whilst the second aims at correcting the deformity itself, the modern tendency is to prefer the latter.

Embryology

The chief development of the heart takes place between the 21st and the 50th day of fœtal life: so that anomalies may be divided into two main types, namely those which occur whilst the heart is forming and those which occur later in intrauterine life. These may be called *failures of development* and *failures after development*.

Failures of Development. The causes of these congenital deformities are poorly understood but experimental and clinical evidence suggest that temperature, nutrition, mechanical and chemical injury, infection of the fœtus, placenta, or the mother, genetic abnormality and parental age may all play some part.

The first evidence of a cardiovascular system is the development of a blood tube

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secundum. Within a few minutes of birth the ductus arteriosus is physiologically closed by contraction of its muscular walls and subsequently by œdema and fibrosis of the muscle and connective tissue, and the circulation becomes adult in form.

The final anatomical obliteration of the ductus arteriosus takes days or weeks, and complete anatomical closure of the foramen ovale is also delayed: and in about 20 per cent of adult hearts it is never complete and a small hiatus persists in the interatrial septum.

The fœtal heart can function normally in spite of anatomical arrangements which would lead to gross abnormality in the adult because there is an extracorporeal circulation through the placenta on which respiration depends. When this extracorporeal respiration ceases at birth the structures adapted to it are obliterated. If they fail to obliterate then blood spills from the greater circulation into the lesser circulation and causes overload and eventual hypertrophy of the right heart and pulmonary vessels.

The Pericardium

The pericardium consists of two parts, a relatively thick, poorly distensible, fibrous sac which surrounds the heart and is attached to the great vessels and parts of the atria, and is also attached to the diaphragm and mediastinal structures; and a serosal portion which invests the heart and lines the inside of the fibrous sac. The fibrous part supports the heart in position, prevents twisting and consequent kinking of the venæ cavæ, and gives attachment for muscular activity; it also helps determine the shape of the heart and by resisting distension limits the volume of the heart. The serosal part allows the heart to move within the pericardial sac with minimal friction. When the fibrous pericardium becomes tense with the larger heart volumes of exercise the mouths of the superior and inferior venæ cavæ are pulled widely open by the fibrous pericardial attachments to these vessels, this effect allows a larger portal of entry of blood into the right atrium.

At rest the normal heart has a diastole volume of about 700 ml., the muscle mass is 300 ml. of this, leaving a blood volume of 400 ml. in the heart. The cardiac output is 140 ml. (70 ml. per ventricle) per contraction which leaves some 260 ml. of blood in the heart at the end of systole; 160 ml. of this is in the atria, and 100 ml. in the ventricles (50 ml. per ventricle). It is thought that the poor distensibility of the fibrous pericardium may lead to certain abnormalities. When the residual blood of one chamber, say the left ventricle, is rapidly increased, as in myocardial infarction, the residual blood in the other chambers is reduced and circulatory abnormalities such as raised venous pressure in the neck and pulmonary œdema occur.

Nevertheless partial or complete congenital absence of the pericardium occurs without abnormality of the circulation.

An undoubted and demonstrable result of the poor distensibility of the fibrous pericardium is the rise in intrapericardial pressure and intracardiac diastole pressure which occurs with accumulation of liquid in the sac (tamponade). A similar rise in intrapericardial and intracardiac diastolic pressure results when the pericardial sac becomes inflamed, fibrous, or calcified as in Pick's disease.

The normal fibrous pericardium may be extensively removed at operation as in pneumonectomy for bronchial carcinoma without causing circulatory upset. It is opened in intracardiac operations and points of practical importance concern the possible dangers

which passes from the caudal to the cephalic end of the embryo, straight across the primitive pericardium, and lying immediately anterior to the notocord. As this tube grows in length it becomes too long for the pericardial sac to contain it, so that an "S" shaped bend forms. The folds of this bend are approximated and differentiate into the various adult chambers of the heart. At the same time as these complicated evolutions are taking place the whole of the systemic vascular network is forming and the system of gill-arches is giving way to the development of lung buds and a new or secondary circulation, the pulmonary system which is formed from modifications of the sixth branchial arch, comes into being. As one contemplates these amazing growths and changes it is strange that most hearts are normal. The following types of anomaly can occur.

(1) *Failure of the heart to descend* into the thorax from the neck; and failure of the sternum to close over it, if already in the thorax. Abnormalities of the pericardium are in this category.

(2) *Aberrations of rotation* such as situs inversus viscerum which may be complete or incomplete and is often symptomless. Dextrocardia in which the heart alone is transposed and which is usually associated with other serious anomalies. Transposition of the aortic and pulmonary trunks which is due to rotation of the aortopulmonary trunk in the wrong direction.

(3) *Persistence of parts of the branchial system* such as double aortic arch, aortic rings, coarctation, etc.

(4) *Failure of the septa to grow* either in part or totally.

(5) *Anomalies of venous return.*

(6) *Various obstructions* within the channels through which the blood should flow, such as pulmonary stenosis.

(7) *Persistence of parts of the fœtal circulation* such as a patent ductus arteriosus.

The failures which occur after the heart has formed, but during the last 7 months of intrauterine life, are very rare and do not concern the surgeon at this moment. They are due to fœtal endocarditis which results in fibrœlastosis, with fusion of the valve cusps, endocardial hypertrophy and secondary myocardial hypertrophy.

Mitral stenosis occurring in the first few years of life is due to a fœtal endocarditis and when it occurs is often associated with a patent interatrial septum (Lutembacher's syndrome).

Fœtal Circulation. In fœtal life the lungs are airless and the pulmonary circulation is small. Most of the oxygenated blood, coming from the placenta through the liver to the heart, passes through the foramen ovale in the interatrial septum into the left atrium and on through the left ventricle into the aorta. Deoxygenated blood coming from the body of the fœtus passes through the tricuspid valve into the right ventricle (the oxygenated and deoxygenated blood streams are kept largely separated in the right atrium) and on into the pulmonary artery, a smaller proportion of this blood passes through the lungs and reaches the left atrium, a larger proportion passes through the ductus arteriosus into the aorta and so reaches the placenta. Pulmonary arterial pressure is therefore higher than aortic pressure and the right ventricle can be considered as the main pump of the placental circulation.

With the first breath the lungs expand, the pulmonary artery resistance falls, more blood flows through the lungs into the left atrium than before, left atrial pressure rises and causes functional closure of the foramen ovale by opposing the septum primum and

Contracting heart muscle fibres are completely refractory to further stimuli, no matter how powerful (absolute refractory period).

Heart muscle fibres also only slowly regain sensitivity to stimuli; early in diastole a strong stimulus causes contraction, late in diastole a weaker stimulus causes contraction (relative refractory period). The practical result of these is that the heart cannot go into tetanus, and after an ectopic stimulus to contraction the long relative refractory period ensures a return to normal rhythm under stimulus from the normal pacemaker.

As regards healing after a surgical incision the muscle of the heart behaves as does muscle elsewhere, namely it heals by replacement of muscle fibre with fibrous tissue. But because muscle is essential for the pump action of the heart wide areas of muscle loss from ischæmia or trauma cause heart failure. In addition, because the cardiac impulse is transmitted through muscle tissue, such injuries may lead at times to blockage of the impulse resulting in partial or total heart block or even asystole.

When the heart beats rapidly the duration of diastole shortens, while the time of systole shortens much less. The result is that with rapid heart rates filling time of the ventricles is much reduced, e.g. when the heart rate rises from 60–140 the filling time is reduced by about 15 seconds per minute.

Automatism of Heart Muscle

Special heart muscle fibres have inherent rhythm; this is more rapid in the SA node (40–150 beats per minute) than in the AV node (30–50 per minute) which is faster than the bundle of His (18–30 per minute).

If the higher parts of this special system become damaged, or isolated from the lower parts, the lower parts act as pacemaker of the common muscle fibres but at slower than the normal rates, e.g. when the bundle of His originates the rhythm in total heart block the heart rate is usually about 30 beats per minute.

The surgeon must distinguish between those movements of the heart which actually pump blood onwards, and those contractions (at times apparently effective) which are completely ineffective.

Pacemaker and Conduction of the Heart Beat

The heart beat normally originates in the pacemaker—the sino-auricular nodal tissue—from here the impulse spreads through auricular muscle in all directions (1000 cm. second) and reaches the AV nodal tissue (the rate of spread is slower in this tissue 200 cm. second) and passes into the bundle of His and on into the branches of this structure (500 cm. second) which lie in two bundles, one on either side of the interventricular septum, and on into further divisions of the system (Purkinje tissue 4000 cm. second) which lie successively under the endocardium of the interventricular grooves, the lateral walls and the base of the heart; and so reach the common muscle cells in all these regions.

AV conduction only takes place via the pathway outlined and the time taken can be measured and corresponds to the PR interval of the electrocardiogram (0·12–0·2 seconds). Rarely an aberrant and shorter band of special tissue connects the auricle and ventricle, the bundle of Kent, and then conduction time is abnormally short (less than 0·1 second).

The sinoauricular node lies in an area to the left of the sulcus terminalis of the right atrium and in the anterior and posterior walls of the right auricular appendix. If the

of leaving the pericardial sac open so that the heart may herniate through the hole, and then undergo torsion with consequent impairment of filling and diminution in cardiac output. On the other hand tamponade will occur if the perciardial sac is tightly sutured and liquid accumulates within it following operation. The post-operative progress after cardiac operations is not always easy, the heart is often larger than before operation, and the difficulty of having to assess the possible part that a pericardial tamponade plays in the clinical picture is an additional complication. The surgeon will therefore prevent herniation, torsion, and tamponade.

The Heart as a Pump

The heart pump needs energy, supplied through the coronary arteries, and anything which diminishes the supply of energy impairs the pump. Anæmia, hæmorrhages, shock, deoxygenation, cardiac tamponade, acute and chronic coronary artery disease all diminish the supply of energy. But the heart cannot be considered alone without creating a meaningless abstraction, for although its sole function is to pump blood it can only do so if the orifices of exit are unobstructed and there is a venous return; and this venous return connects it through the veins, venules, capillary pool, arterioles, and arteries, which are all controlled by nervous impulses and hormones, with the cardiac output.

The heart may cease to function efficiently as a pump, during the course of an operation, for any of the following reasons. The volume of blood available becomes inadequate to fill the pump, the energy source of the pump fails, the channels through which blood should pass are obstructed, the myocardium is poisoned, or the respiratory and vasomotor centres in the brain are injured.

It is the surgeon's business to see that these qualities are not impaired by his manipulations.

Muscle Structure and Properties

Heart muscle is of two sorts, specific conduction tissue and common muscle. The specific tissue is present in the SA node, the AV node, the bundle of His and its major branches, the terminal divisions and subdivisions of which lie mainly beneath the endocardium of the ventricle but also within the ventricular wall. The ventricular portion of this special tissue can be visualized as a fine sponge with the interstices filled up with common muscle. It follows from this that the surgeon who operates on the heart must know the exact anatomical situation of the larger masses of conduction tissue, SA node, AV node, bundle of His, and main branch bundles, because any injury to these may lead to conduction defects causing heart block of varying degree. The finer ramifications of the conduction system are not so important; some must be divided by any incision of the ventricular muscle and no abnormality of conduction results.

Heart muscle has certain characteristics peculiar to it: it is excitable, it contracts, the contraction is conducted through the muscle mass, and the contractions are rhythmic. Heart muscle fibres are interconnected by bridges of tissue making a whole. Skeletal muscle fibres are insulated and individually separate. One skeletal muscle fibre can contract without any others contracting. In the heart because there are no isolated muscle fibres, the contraction, wherever it originates, spreads through all the fibres. Skeletal muscle fibres can be stimulated to contract again while in a state of contraction and if sufficiently rapidly stimulated will go into a state of continuous contraction (tetanus).

Digitalis is the specific drug of choice for the *control* of these three abnormalities, auricular fibrillation, auricular flutter and paroxysmal auricular tachycardia, while quinidine may be used to *abolish* the abnormality.

Paroxysmal Ventricular Tachycardia. In this condition the function of the normal pacemaker is usurped by an ectopic focus in the ventricle, the rate is rapid and regular but not quite so absolutely unvaryingly regular as it is in paroxysmal auricular tachycardia.

Ventricular paroxysmal tachycardia is almost always associated with grave myocardial disorder and is a common prelude to ventricular fibrillation. The prognosis is bad whereas paroxysmal auricular tachycardia is benign. The accurate differentiation

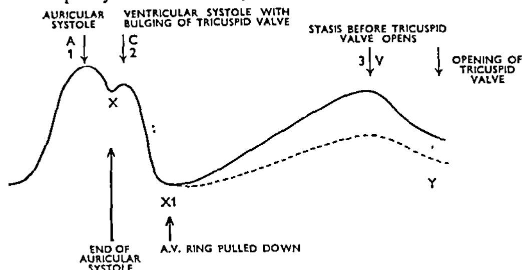


FIG 259. Diagram of the movements visible in the great veins in the neck with the causes of the peaks and troughs indicated.

of these conditions rests in the E.C.G. pattern which must be obtained because treatment is different. Paroxysmal ventricular tachycardia should be treated by drugs other than digitalis as for example quinidine sulphate or procaine amide (Pronestyl).

Heart Block. Total heart block is a complete impulse dissociation between atria and ventricles, in which the ventricular rate depends on the rhythmicity inherent in the ventricular conduction system—usually 20–40 beats per minute. This inherent rhythmicity is feebler than atrial rhythmicity and there is a constant tendency for ventricular standstill with the resulting cerebral anæmia, anoxia, and epileptic convulsions. Total heart block is a grave condition and sudden death is not rare. When it is present some interference with the main bundle of His can be inferred; commonly myocardial disease. The surgeon who approaches the heart must avoid manœuvres which encroach on the bundle of His and so cause total heart block.

A common condition is blockage of conduction in one or other branch of the bundle of His which produces abnormal electrocardiographic patterns. It is important because its presence may indicate unbalanced hypertrophy, a strain of one ventricle while the other remains normal, e.g. right ventricular hypertrophy in pulmonary stenosis, or when there is pulmonary embolism, and left ventricular strain and hypertrophy in systemic hypertension. Local disease in one bundle may also produce this E.C.G. pattern.

The clinical importance of this form of heart block is the clear cut evidence it affords that the myocardium of one ventricle is under abnormal stress and is responding unfavourably, e.g. in pulmonary or aortic valve stenosis the finding of a bundle branch

sulcus terminalis is mistaken for part of the rim of an atrial septal defect, not a rare mistake, then this structure may be involved in sutures.

The auriculo-ventricular node lies in the lower part of the interatrial septum just above the point of attachment of the septal cusp of the tricuspid valve, and near the orifice of the coronary sinus. The main conducting bundle of His runs from this point across the auricular-ventricular ring to descend along the lower margin of the membranous interventricular septum and enter the muscular septum where it divides into right and left branches which descend and spread fanwise over the inner surfaces of the ventricular walls.

It will be seen that operations to close atrial and ventricular septal defects will be performed close to these vital structures and they must not be involved in sutures or otherwise injured.

Abnormalities of Heart Rhythm

Auricular Fibrillation. When the myocardium is diseased or ischæmic or hypertrophied for any reason, such as valve disease or hypertension, the normal pacemaker system (A.V. node) and normal impulse propagation may become deranged. Auricular fibrillation is one of the abnormalities which may occur. In this the impulse, instead of arising in the S-A node and spreading evenly, ripplewise, through both atria to reach the A-V node, is generated in some ectopic site in the atria. The impulse formation is abnormally rapid (450–600 impulses per minute) and spreads unevenly through the atrial wall; as a result impulses reach the A-V node more often than it can respond, and also irregularly in time. The A-V node initiates ventricular contractions as often as it can respond to these rapid and irregular stimuli, but the contractions are irregular in rhythm and result in irregular ventricular stroke outputs of blood because filling times are unequal. As a result although the heart beats abnormally rapidly the cardiac output may diminish below normal rather than be increased as it is in physiological tachycardia. Heart failure results; blood tends to stagnate in the uncontracting atrial appendages and thrombi form, and these may be thrown as emboli into the greater or lesser circulation.

Although experimental and some small amount of clinical evidence supports the concept of uniatrial fibrillation, for all practical purposes auricular fibrillation may be considered as affecting both atria.

Auricular Flutter. In auricular flutter which may occur in conditions similar to those which cause auricular fibrillation, the ectopic atrial impulse formation is regular and rapid (about 300 impulses per minute) but not so rapid as in auricular fibrillation.

The ventricle responds as frequently as the refractory period allows—commonly one-half or one-third as often as the auricular rate of impulse formation (150 beats per minute). A sometimes useful clinical diagnostic point is this: vagal stimulation may suddenly halve the rate of ventricular response (i.e. 150 beats to 75 beats per minute) during the time the stimulus is applied, but the ventricular rate will double back to the original rate as soon as the stimulus is removed. In paroxysmal auricular tachycardia such stimulation may permanently stop the tachycardia and induce normal atrial impulse formation.

Paroxysmal Auricular Tachycardia. This is commonly a benign condition but it may also occur with heart disease. Ectopic impulse formation at regular and rapid rate occurs (usually about 200 impulses per minute) and the ventricle responds at the same rate.

Digitalis is the specific drug of choice for the *control* of these three abnormalities, auricular fibrillation, auricular flutter and paroxysmal auricular tachycardia, while quinidine may be used to *abolish* the abnormality.

Paroxysmal Ventricular Tachycardia. In this condition the function of the normal pacemaker is usurped by an ectopic focus in the ventricle, the rate is rapid and regular but not quite so absolutely unvaryingly regular as it is in paroxysmal auricular tachycardia.

Ventricular paroxysmal tachycardia is almost always associated with grave myocardial disorder and is a common prelude to ventricular fibrillation. The prognosis is bad whereas paroxysmal auricular tachycardia is benign. The accurate differentiation

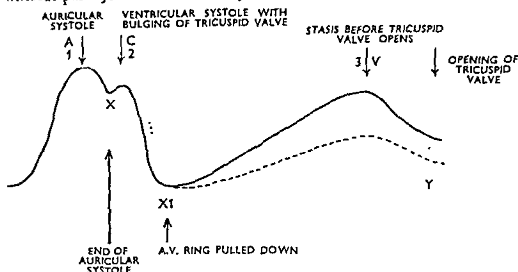


FIG 259. Diagram of the movements visible in the great veins in the neck with the causes of the peaks and troughs indicated.

of these conditions rests in the E.C.G. pattern which must be obtained because treatment is different. Paroxysmal ventricular tachycardia should be treated by drugs other than digitalis as for example quinidine sulphate or procaine amide (Pronestyl).

Heart Block. Total heart block is a complete impulse dissociation between atria and ventricles, in which the ventricular rate depends on the rhythmicity inherent in the ventricular conduction system—usually 20–40 beats per minute. This inherent rhythmicity is feebler than atrial rhythmicity and there is a constant tendency for ventricular standstill with the resulting cerebral anæmia, anoxia, and epileptic convulsions. Total heart block is a grave condition and sudden death is not rare. When it is present some interference with the main bundle of His can be inferred; commonly myocardial disease. The surgeon who approaches the heart must avoid manœuvres which encroach on the bundle of His and so cause total heart block.

A common condition is blockage of conduction in one or other branch of the bundle of His which produces abnormal electrocardiographic patterns. It is important because its presence may indicate unbalanced hypertrophy, a strain of one ventricle while the other remains normal, e.g. right ventricular hypertrophy in pulmonary stenosis, or when there is pulmonary embolism, and left ventricular strain and hypertrophy in systemic hypertension. Local disease in one bundle may also produce this E.C.G. pattern.

The clinical importance of this form of heart block is the clear cut evidence it affords that the myocardium of one ventricle is under abnormal stress and is responding unfavourably, e.g. in pulmonary or aortic valve stenosis the finding of a bundle branch

block may be an indication for valvotomy, while it also indicates the dangers of such a manœuvre.

Electrical Activity of the Myocardium

Muscle cell activity generates electrical current. A heart muscle cell may be regarded as a sphere. At rest it is polarized, that is the cell membrane is positively charged and the interior of the cell is equally but negatively charged, in this state the cell membrane is impermeable and no current flows. If any part of the cell surface is stimulated it becomes permeable, negatively charged ions flow out through the membrane (depolarization) and an electrical current is produced which stimulates the cell surface adjacent to the original point stimulated and progressive depolarization occurs. The quantity of current increases till half the surface area of the cell membrane is depolarized and then decreases until the whole surface is depolarized. The repolarization starts immediately, at the point originally stimulated and spreads from this point till the whole of the cell membrane is repolarized; this repolarization is a rather slower process than depolarization and while it is occurring current flows in a reverse direction to the current of depolarization.

These currents which originate within the body flow in three dimensions throughout it, but less freely in some parts than in others; and the currents can be recorded and form the basis of the electrocardiogram.

The practical usefulness of the electrocardiogram during operation is that it gives an immediate record of the electrical activity of the myocardium and may therefore warn that anæsthesia, anoxia, cold, manipulation, etc., are causing damage. The surgeon will also note that the electrocardiogram may appear normal when the heart is already an inadequate pump for the existence of electrical activity only reports that muscular contraction is occurring and tells nothing of the results of contraction: e.g. the heart continues to beat and produces a normal electrocardiogram when it is isolated and an extracorporeal pump maintains the circulation.

No systematic description will be given of electrocardiography on which there are numerous texts to which the reader is referred. But where the electrocardiogram is useful in the diagnosis of a specific abnormality a brief description will be given.

Atrial Activity

It seems from both practical and theoretical considerations that atrial systole is of no great importance in ventricular filling and cardiac output. Prinzmetal believes that right atrial activity is greater than left atrial activity, and also that in exercise the right atrial systole plays an important part in ventricular filling. The relative insignificance of atrial systole may also be inferred from the fact that patients with atrial standstill may be otherwise normal, and also that patients with "idiopathic" auricular fibrillation under control, in whom the heart is otherwise apparently normal, can be normally active. It seems clear that the atria serve as sizeable reservoirs from which large quantities of blood can rapidly empty into the ventricles either wholly passively or with a small assistance from auricular contraction.

The fact that atrial systole only occupies $\frac{1}{8}$th of the ventricular diastole (0·05 sec. of 0·4 sec.) also suggests that it is of relatively small importance in ventricular filling, except when the heart rate is rapid and atrial systole occupies a large proportion of ventricular diastole, and also when mitral or tricuspid valve obstruction interferes with

ventricular filling the atrial hypertrophy which results makes atrial activity an important factor in ventricular filling.

An important result of atrial hypertrophy is the eventual failure of co-ordinated auricular contraction which results in auricular fibrillation with stagnation of blood, particularly in the atrial appendages, with thrombus formation and embolization of the lungs from the right atrium and of the systemic arteries from the left atrium. It is estimated that patients with mitral stenosis and auricular fibrillation are six times more liable to emboli than patients with mitral stenosis in normal rhythm.

Metabolism

Heart muscle normally obtains energy for contraction by the metabolism of lactic acid, and also to a small extent in emergency by the metabolism of glycogen. But it appears that heart muscle is versatile and under abnormal conditions fat can be used for a short while before contraction becomes abnormal and stops. In addition to a source of energy, normal temperature (37°C.), pH, suitable oxygen supply, calcium, potassium, and sodium in adequate concentrations are all necessary for normal contraction, relaxation, normal rate, and normal output. Experimentally, proteins are apparently not important. Vitamins are essential, heart muscle requires Vitamin B complex (Theamin, Nicotinamide, and Riboflavin) in order to metabolize carbohydrate, and there is evidence that Vitamin E appears to be necessary for normal action.

With temperatures between 26°C.–40°C. the heart still beats, but either abnormally slowly or abnormally fast; with the lower range of temperature auricular fibrillation, auricular standstill and impulse conduction block may develop.

Increased acidity (low pH) causes progressively greater relaxation till the heart stops in diastole; increased CO_2 has the same effect and also dilates the coronary arteries but this latter effect is opposed by central vaso-constriction. It will be noted that the blood CO_2 and lactic acid accumulation due to exercise will relax the heart more completely and permit greater filling and it may also have some effect in increasing coronary flow.

Alkalis shorten diastole and lengthen systole and as pH rises impulse conduction is impaired, heart block develops and the heart stops in systole. Alkalis have no effect on the coronary flow. Heart muscle must receive an adequate supply of oxygen and only tolerates short periods of oxygen lack. Heart muscle normally extracts a high proportion of oxygen from the circulating blood.

High concentrations of oxygen reduces the heart rate, and cardiac output; low concentrations increase the rate and output. Very low 0_2 concentrations cause premature beats, arrhythmia and heart block and finally heart failure and cardiac arrest. Low concentrations diminish the height of the T wave and cause T wave inversions like those of myocardial infarction. Breathing low tension 0_2 has been used as the basis of tests of coronary artery disease.

Calcium, potassium, and sodium ions in suitable proportions are essential to normal heart action. Calcium ions if present either alone or in excess cause increased length of systole and finally the heart stops, contracted in systole (calcium rigor). Potassium either alone or in excess relaxes the heart and causes it to stop relaxed in diastole. (Potassium inhibition.) Highly characteristic and diagnostic changes occur in the E.C.G. consisting of tall and sharply peaked T waves when the serum potassium rises above the normal.

block may be an indication for valvotomy, while it also indicates the dangers of such a manœuvre.

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very rapid heart rates which diminish diastole, without a corresponding increased venous return such as occurs in exercise, diminish coronary flow.

The coronary arteries have both sympathetic and parasympathetic nervous control, the former through the upper six thoracic sympathetic ganglia, the latter through the vagus nerves; the sympathetic is vasodilator, the parasympathetic vasoconstrictor. Stimulation of the gall bladder, bile ducts, œsophagus, and stomach may all cause reflex vasoconstriction mediated through the parasympathetic.

Drug Effects on Coronary Artery Flow

Direct dilation of the coronary arteries with increased flow is caused by anoxia and asphyxia and also by various drugs, among which are:

Adrenaline.
Alcohol.
Aminophylline.
Ephedrine.
Khellin (Amini Visnaga).
Nitrites.
Papaverine.
Theophyllin.

Morphia increases flow in an unknown way, possibly by a central effect.

Tachycardia and raised blood pressure which result from thyroid hormone and insulin increase coronary flow.

Atropine increases flow by vasoconstrictor inhibition.

Digitalis in therapeutic doses increases flow by increasing the cardiac output and slowing the rate.

Coramine increases flow.

Direct vasoconstriction of the coronary arteries with decreased flow is caused by foreign protein shock, nicotine, histamine, pituitrin, pitressin, and angiotonia.

Acetylcholine, Mecholyl, and Doryl by vagal action decrease coronary flow.

Digitalis in toxic doses, possibly as a result of vagus inhibition and the tachycardia it produces, decreases coronary flow.

Coronary Veins

The veins of the heart are extensive and consist of several systems which inter-communicate within the muscle mass of the various chambers of the heart. These venous systems are the coronary sinus which enters the right atrium, the lesser veins which enter the right atrium, Thebesian veins which open directly into all the heart chambers and the intramuscular sinusoids (the arteriolumenal and arteriosinusoidal) which drain into the heart cavities.

Experiment has shown that the coronary sinus can be made effectively to carry arterial blood into the myocardium after suitable measures have been taken to increase the capacity of the communications between the coronary sinus and the other cardiac venous drains.

Further it has been shown that an open artery (the internal mammary—intercostal)

Sodium is essential to continued rhythmicity of the heart, and without sodium the heart stops contracting after a while. Alkalis behave like calcium, acids behave like potassium.

Hyperkapnia is dangerous. It causes the serum potassium to rise and if subsequently sufficient oxygen is given the potassium level rises still further and ventricular fibrillation with circulatory arrest may occur; and there is little doubt that a raised serum potassium is more dangerous still when there is hyperkapnia. Hence the importance of careful anæsthesia. A continuous E.C.G. trace is valuable in detecting this, for as the potassium rises the T waves become characteristically tall and sharply pointed; if this is seen to occur, 3 per cent NaCl and 20 per cent glucose should be given at once intravenously.

It follows that precautions must be taken to prevent pathological alterations in the blood during cardiac operations, particularly when an extracorporeal circulation is used; and perfusion of the coronaries with citrated blood during an operative emergency is likely to prolong cardiac arrest. The heart may be stopped during certain intracardiac operations, done under hypothermia, by perfusing the coronary circulation with potassium salts. This type of "controlled arrest" is useful to the surgeon not only because the parts are still, flaccid (as opposed to distended with blood) and easily manipulated, but the period of interruptions of the circulation can be increased because the myocardium is not using much oxygen or accumulating much metabolite. Normal rhythm can be started again after the "controlled arrest" by perfusing the coronaries with blood containing calcium salts.

Coronary Artery Circulation

The coronary circulation supplies the heart muscle with oxygen and nutritive material (lactic acid, glucose, fat). Any impairment in this circulation, whether stoppage of flow, or diminution of essential materials, will impair contraction. Circulation depends on sufficient gradient; liquids flow from high pressure regions to lower pressure regions, and the absolute pressure is of lesser importance. If the periphery is widely open an adequate flow will take place with a small absolute pressure. Examples of this may be seen daily with modern hypotensive drugs in anæsthesia: the blood pressure is low but the flow is adequate because the peripheral resistance to flow falls. The volume of blood flow is therefore all-important and what matters is that an adequate quantity of oxygen and nutritive material arrives each second or minute or hour. For short periods of time, a few minutes at most, the heart muscle can tolerate famine. In this respect skeletal muscle is much more adaptable, it can contract a five times larger O_2 debt than heart muscle, and athletes' muscles can be trained to borrow more than sedentary muscles Heart muscle has only a small capacity for working in this way and must pay as it goes or it soon becomes insolvent and fails

Anoxia is the greatest common danger to myocardial activity and it is immediately caused by anything which interferes with the coronary circulation. Even a small bubble of air in a coronary artery may lead to anoxia of a portion of myocardium with the risk of ventricular fibrillation

The coronary artery circulation is some 5 per cent of the cardiac output at rest (76 ml. per minute per kg. body weight is an accurate formula). Blood flows both during systole and diastole, with a larger quantity and higher velocity during diastole. For this reason

Anyone can train himself to observe the height of the venous wave in the neck and its characteristic form and also *the variations from normal* which occur in disease. A close study of venous tracings and the observed movements in the neck can result in a high degree of accuracy.

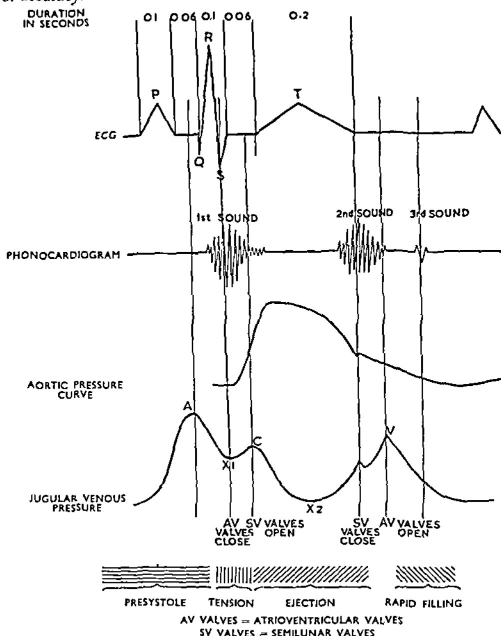


FIG 260. Diagram to show relationship of jugular venous pressure waves to simultaneous electrical, auditory, and mechanical events.

Cardiac Output

To measure the cardiac output the direct Fick principle is employed as convenient and reasonably accurate. The postulate is that if the oxygen content of blood entering the lungs is known, and the oxygen content of blood leaving the lungs is known, and if a simultaneous measurement of the quantity of oxygen taken up by the blood in passage through the lungs is made then the quantity of blood which has flowed through the lungs

can be implanted directly into the myocardium and no hæmatoma results though the artery remains open and pumps arterial blood into the myocardium. It appears from this that the sinusoidal venous drainage system in the myocardium is capable of carrying off large volumes of blood. There are valves guarding the origins of the tributary veins of the coronary sinus and the entrance of the coronary sinus into the right antrium, but it is clear from experimental work in animals and man that when the drainage of blood from the coronary sinus into the right atrium is interrupted by ligature the valves of the tributary veins become incompetent and blood flow is reversed, and also, when the coronary sinus is anastomosed with an artery, blood flows retrograde in the tributaries, the guard valves of which become incompetent owing to distension.

The Venous Pulse

Observation of the venous pulsation in the neck is important in the diagnosis of heart disease. Venous pulsation may be abnormal in two ways, either the level at which the pulsation occurs is abnormally high in the neck, or the typical wave form is altered.

Abnormally high venous pressure is evidence of heart failure or of obstructed venous return, and the latter can be distinguished because the characteristic pulsations are damped.

Venous movement can be separated from arterial movement, the former is multiple during each cardiac cycle, the other is single. Furthermore venous movement can be more easily seen than felt with the finger, and any movement which cannot be felt may be taken to be venous, though venous movement such as occurs with an hypertrophied right atrium or with tricuspid incompetence may occasionally be palpable.

The venous pulse in the neck has a characteristic form (Fig. 260). Descriptive letters are applied to the various elevations and depressions of the wave. These will be described because an understanding of the wave form is essential. The events described take place in the right atrium but similar events occur in the left atrium and a venous wave is present in the pulmonary veins slightly later in time because the left atrium is excited to contract very slightly after the right auricle (0·013 sec. delay).

(1) A wave—auricular systole raises intra-auricular pressure and produces this wave. In auricular fibrillation this wave disappears, and in conditions in which the right atrium is hypertrophied this wave will become abnormally larger.

(2) C wave—ventricular systole not only closes the tricuspid valve but it causes the cusps to bulge into the right atrial chamber and raises the pressure there and produces this wave.

(3) V wave—following ventricular systole and before the tricuspid valve opens blood collects in the right atrium and great veins, the pressure rises to produce the V wave, the tricuspid valve opens and the pressure falls.

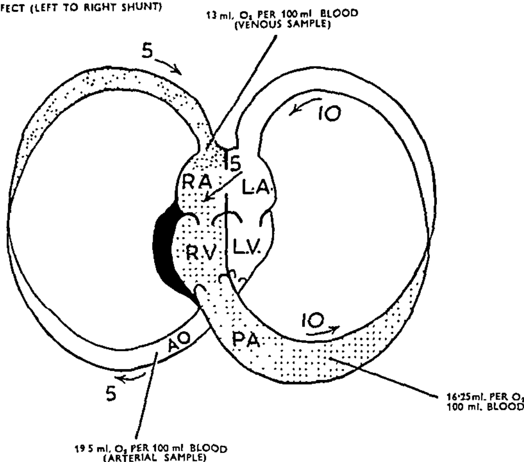
The troughs or depressions are produced by these events.

(1) X depression—caused by the drop in intra-auricular pressure at the end of auricular systole.

(2) X_1 depression—the earliest movement of ventricular systole is a shortening of the interventricular septum, this pulls the AV ring down towards the ventricular apex and this by enlarging left auricular capacity causes the pressure to fall.

(3) Y depression—caused by opening of the tricuspid valve when blood flows out of the right atrium and pressure falls.

(3) Oxygen absorbed in lungs = 325 ml. O_2 per minute. Then A-P = 3·25 ml. O_2 which has been taken up by each 100 ml. of blood in passage through the lungs. Therefore 325 ml. O_2 absorbed in the lungs represents a passage of $\frac{325}{3 \cdot 25} \times 100$ ml. = 10,000 ml. blood per minute through the lungs. But this flow *includes a quantity of already fully oxygenated blood* which has been shunted through the atrial septal defect from the left



NOTE The small figures represent flow of blood in litres per minute

FIG 262 Cardiac output in a patient suffering from an atrial septal defect and having a left to right shunt.

atrium. Clearly this part of blood has taken up no oxygen in its further passage through the lungs as it is already saturated.

Now if the flow is calculated from the mixed venous and arterial samples as before it will be found that 5,000 ml. of deoxygenated blood (13 vols. per cent) have flowed through the lungs. Therefore as the previous calculations gave a flow of 10,000 ml. 5,000 ml. of this must be O_2 saturated blood, i.e. the atrial septal defect has shunted 5,000 ml. blood per minute and 5,000 ml. have gone into the greater circulation.

In this case the systemic flow is 5,000 ml. per minute and the pulmonary flow is 10,000 ml. per minute. Right ventricular hypertrophy must result from this overwork. The same observations apply when there is a large ventricular septal defect or a large patent ductus arteriosus

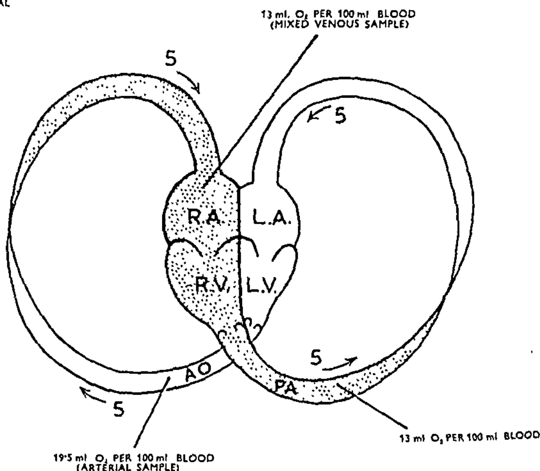
an be calculated. If there are no intracardiac shunts this quantity is the cardiac output.

For example: (in Fig. 261).

(1) Mixed venous blood = 13 ml. O_2 per 100 ml. blood (V).

(2) Arterial blood = 19·5 ml. O_2 per 100 ml. blood (A).

(3) Oxygen absorbed in lungs = 325 ml. O_2 per minute. Then A-V = 6·5 ml. O_2 which has been taken up by each 100 ml. of blood in passage through the lungs. Therefore 325 ml. O_2 absorbed represents a passage of $\frac{325}{6 \cdot 5} \times 100$ ml. blood per minute = 5,000 ml. blood per minute through the lungs. As there is no shunt this represents the cardiac output and flow of blood through the greater and lesser circulations.



NOTE The small figures represent flow of blood in litres per minute

FIG 261 Cardiac output in the normal

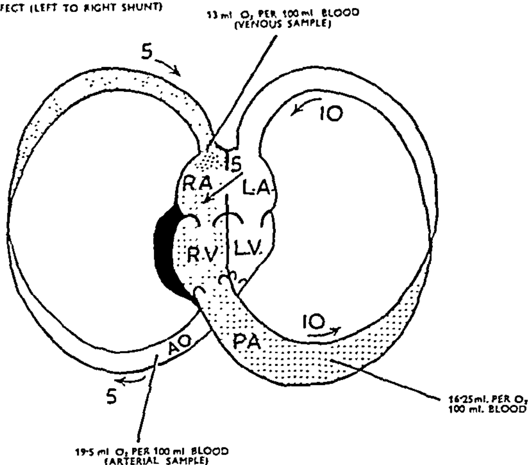
In the same way if there is an intracardiac shunt from left to right side, as when there is an atrial septal defect or a ventricular septal defect or a patent ductus arteriosus then a measurement of the pulmonary blood flow and a separate measurement of the systemic blood flow can be made if the oxygen content of the mixed venous blood, the pulmonary artery blood and the arterial blood, and the oxygen uptake in the lungs are known.

For example: (in Fig. 262)

(1) Pulmonary artery blood = 16·25 ml. O_2 per 100 ml (P).

(2) Pulmonary venous blood (arterial blood) = 19·5 ml. O_2 per 100 ml. (A).

(3) Oxygen absorbed in lungs = 325 ml. O_2 per minute. Then A-P = 3·25 ml. O_2 which has been taken up by each 100 ml. of blood in passage through the lungs. Therefore 325 ml. O_2 absorbed in the lungs represents a passage of $\frac{325}{3 \cdot 25} \times 100$ ml. = 10,000 ml. blood per minute through the lungs. But this flow *includes a quantity of already fully oxygenated blood* which has been shunted through the atrial septal defect from the left



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FIG 262 Cardiac output in a patient suffering from an atrial septal defect and having a left to right shunt.

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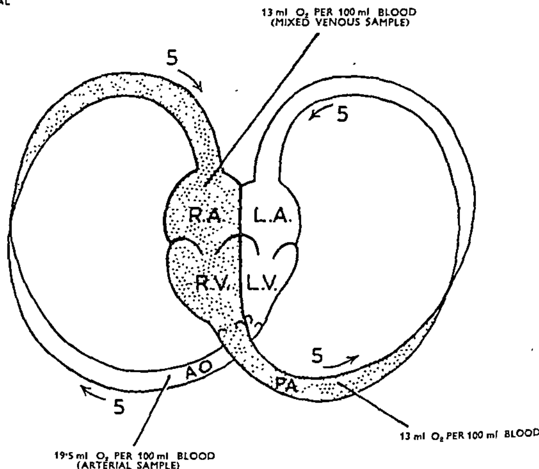
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FIG 264 Early filling (oblique view)



FIG 265. Late filling (A.P. view)



FIG 266 Early filling (A P. view).

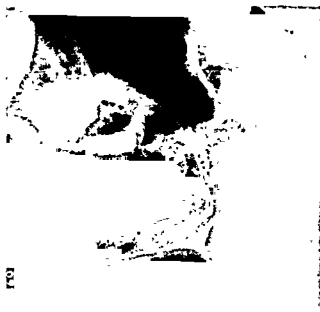


FIG 267. Late filling (oblique view).

The Cardiac Catheter

Intracardiac and Intravascular Pressures. By venous intubation and manometry the pressures within the great veins, the right atrium, the right ventricle, the pulmonary arteries, and the "pulmonary capillaries" may be measured and permanently recorded. The most suitable veins to use for entry are the left and right antecubital veins and if these are for any reason not suitable then the femoral veins, but the latter have the disadvantage that venous thrombosis, if it should occur, may lead to massive pulmonary embolism.

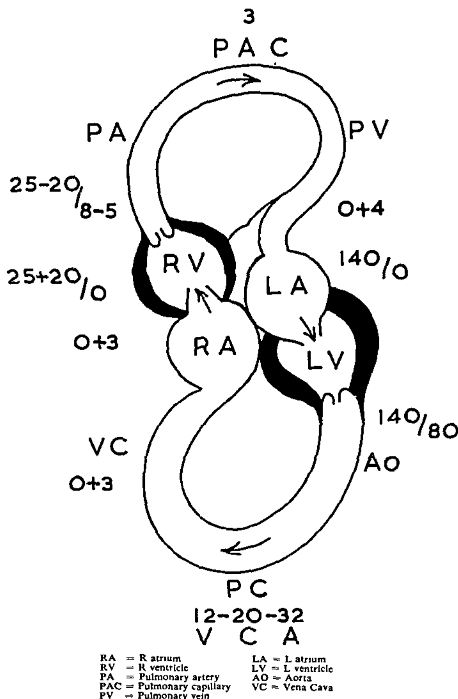


FIG 263 Normal pressures in various parts of the circulation—in millimetres of mercury.

The balloon is placed in the defect and distended with radio-opaque material and its position is then determined.

Intracardiac and Intravascular Blood Samples. In addition to obtaining pressure measurements the cardiac catheter can also be used to obtain samples of blood for gas analysis from the various chambers of the heart and from the great vessels.

The exact position of the tip of the radio-opaque catheter can be checked by direct fluoroscopy, and a further check on the position of the catheter tip can be made by observing the pressure wave form, suitably recorded, which is characteristic in each chamber.

Dangers of Cardiac Catheterization. The dangers inherent in these manœuvres is small in experienced hands. Failure to pass the catheter because of venospasm can be prevented by neat and rapid introduction. Thrombosis can be controlled by anti-coagulants. Cardiac arrhythmias are not rare and are due to irritation of the heart by stimulation of the endocardium; ventricular and auricular extrasystoles are commonest, but auricular tachycardia is not rare and such arrhythmias will probably interrupt the procedure. When the patient is on the verge of acute pulmonary œdema the excitement and fearfulness produced by the manœuvre may be sufficient to cause dangerous and even fatal pulmonary œdema. Rarer mishaps may occur such as knotting of the catheter inside the heart, and very rarely there may be perforation of the heart. Embolus following cardiac catheterization is not common.

Remember that misleading results may occasionally be obtained in the blood samples for gas analysis, because the blood in the right atrium tends to flow in streams, i.e. the superior vena caval blood and the inferior venal caval blood are kept partially separate, and in some cases of atrial septal defect the shunted blood may also flow separately. For this reason where there is doubt samples from the venæ cavæ, the right atrium, right ventricles, and pulmonary artery must all be obtained, e.g. even though the right atrial sample suggests no left to right shunt because the shunted stream has not been sampled, then a right ventricular and pulmonary artery sample will show the shunt because the streams mix in the ventricle.

Angiocardiography

The principle is to use a radio-opaque substance in solution of suitable strength, and to inject sufficient material at an adequate speed so that a concentration is achieved in the heart and great vessels. In this way a shadow profile of the various cardiac chambers can be obtained by taking films in rapid succession. Furthermore by observing the sequence of filling, right atrium, right ventricle, pulmonary artery, any deviation from the normal order can be noted and inferences made, e.g. if the aorta is shown to fill immediately after the right ventricle the existence of a right to left shunt can be surmised.

The solution is commonly introduced into the antecubital vein through a wide bore needle or canula, and although this results in some dilution by the blood which lies between the point of injection and the heart this is not sufficient to eliminate the contrast in the films. When the left side of the heart and the aortic root and arch are being investigated the dilution which is due to blood between the site of injection in the arm and the left side of the heart is much greater, including as it does the volume of blood in the lungs, and indifferent contrast is obtained in the films. For this reason when the aorta is to be filled with contrast material it is usual to inject directly into the

The precise technique of cardiac catheterization are not dealt with here. The left side of the heart, i.e. the left atrium, the left ventricle, and the aorta can also be explored by catheter which can be introduced into the left atrium, via a bronchoscope, through a needle passed through the bronchial wall and into the left atrium; an alternative method is to introduce the catheter through a needle which pierces the left atrium directly through the chest wall across the pleural cavity and across the pericardium, When there is mitral stenosis or aortic stenosis if the catheter can be passed through the contracted valve

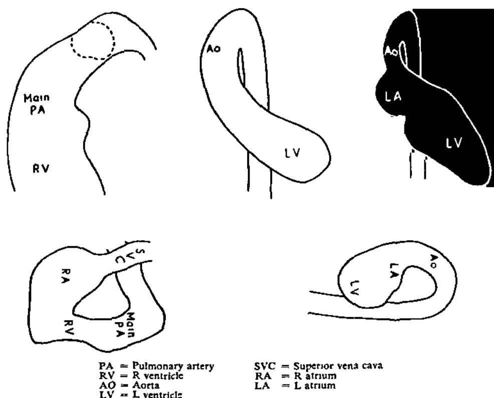


FIG 268 These drawings explain Figs. 260, 261, 262, 263.

orifice then an exact measurement of the pressure difference (gradient) on either side of the valve can be obtained. Similarly by catheterization of the right side of the heart when there is tricuspid stenosis or pulmonary stenosis the pressure differences can be obtained. The discovery of an important pressure difference on either side of a valve may, when taken together with the patient's symptoms and the discovered physical signs, afford clearcut evidence for surgical correction of the abnormality.

The diagram shows the normal pressures found in the adult.

Intracardiac and Intravascular Position of Catheter. At times most useful information may be obtained by watching the catheter under the fluoroscopic screen, e.g. the discovery of an increased right atrial pressure and of oxygenated blood within the right atrium may suggest an atrial septal defect with a predominant left to right shunt of blood; but the catheter may be observed to pass directly into the lung fields thereby proving the existence of anomalous drainage of pulmonary veins into the right atrium.

Intracardiac Anatomical Exploration. The cardiac catheter can also be used, if fitted with a distensible balloon, to determine the size and position of intra-atrial septal defects.

mechanical cause of muscle hypertrophy has been removed early enough a normal life may be expected (remembering always the possible presence of associated anomalies such as intracranial aneurysm, bicuspid aortic valves, aortic cystic medianecrosis.

Pick's disease (constrictive pericarditis) is an example of failure of venous input due to inadequate distensibility of the heart chambers to accommodate the venous return, but there is also an associated muscle failure as the myocardium is always damaged in its outermost layers by the pericardial fibrosis, and sometimes by fibrosis which extends into the deeper layers of muscle, and there is also a possible involvement of the coronary arteries with inadequate nutrition of the myocardium.

The surgeon usually meets heart failure as a sudden inadequacy or stoppage of the heart-beat which results from ventricular fibrillation or ventricular asystole, and formerly this had a catastrophic result even though the heart would sometimes be made to beat again. The physician in contrast knows heart failure as a long-drawn inadequacy of the heart's action which leads to a characteristic syndrome comprising effort dyspnœa, raised venous pressure, œdema, liver enlargement, congestion of the lung bases, pleural effusions and ascites The steps the surgeon should take when he is confronted with ventricular fibrillation or cardiac asystole are described elsewhere (q.v.).

Prognosis in Heart Disease

By far the most difficult fact to establish with certainty in a patient with heart disease is the prognosis; and on what we believe to be the prognosis for life or disability we base our treatment. For instance, even in a condition such as coarctation of the aorta, so intensively studied recently, although it is widely recognized that the average age at death is 33 years this figure nevertheless only offers a very rough guide, for every patient is an individual with the very varying circumstances which apply uniquely in his case. Everyone will recall instances from his own experience where the outcome destroyed the prognosis, and when we consider among the many unponderables the age, sex, duration of illness, morale, economic state, occupation, structural changes, possible complications, response to therapy and so forth this is hardly surprising. This fact of difficult prognosis in heart disease still controls and limits the surgery of the heart. A few observations which may be useful are given.

A quick heart gives a better outlook for therapy. Heart failure which cannot be controlled by medical means is a bad surgical omen

A big heart means a bad prognosis; exceptions are acute rheumatic carditis, vitamin deficiency heart disease, and myxœdema. Although a big heart with mitral valve disease is a strong contraindication to operation in a young person, the same size of heart is not necessarily a contraindication to surgery in older patients, and many have been helped by surgery.

The effects of constrictive pericarditis will disappear if the disease is recognized and dealt with. Success or failure depends upon the myocardium, and to a much lesser extent on the existence of cirrhosis of the liver.

Infections such as acute rheumatism tend to recur and are probably a continuous process throughout life without a termination like that in, say, lobar pneumonia. As a result the good effect of surgery in rheumatic heart disease will sometimes prove to be short-lived and operation will have to be repeated.

aorta. This has been done into the aorta through the chest wall but it is safer to pass a catheter retrograde from the radial or the carotid artery till the tip lies in the ascending aorta and the contrast material is injected there. Similarly it may be an advantage at times when there is great venous engorgement and enlargement of the right atrium and ventricle to inject the medium through a catheter passed into the heart via the antecubital vein. The problems of dilution can in this way be overcome. Special apparatus is necessary to achieve a rapid delivery of material against the normal aortic pressure through a fine bore tube. It should be noted that injecting radio-opaque material in high concentration into the coronary arteries may prove lethal. (Figs. 264, 267.)

The danger from these manœuvres is small.

Heart Failure

The heart fails when it no longer puts out sufficient blood to supply the needs of the body although the venous return is adequate.

If the volume of fluid in the vascular bed (arteries, capillaries, veins) is reduced and vascular tonus is inadequate to maintain the intravascular pressure then the venous return will fall and so will the cardiac output. This may happen in acute hæmorrhage or in shock. This is failure of the circulation but not heart failure. It is important to stress this distinction because the treatment of circulatory failure is to restore intravascular fluid volume and tone before the various organs of the body, including the heart, are damaged by inadequate blood supply. On the other hand where there is failure of heart muscle the heart muscle must be assisted by means such as digitalis and removal of the cause. The cardiac surgeon should recognize that circulatory failure and heart failure may co-exist but are separate entities.

Heart muscle may fail to maintain adequate output for various reasons. There may be acute or chronic coronary artery insufficiency, there may be intoxication, inflammation or degeneration of heart muscle cells, as in diphtheria or rheumatic fever for example, or in the rare amyloid deposition; the muscle failure here is absolute and is not related to any other fault in the circulation. Cure will depend upon reversal of the process which may be possible in diphtheria and rheumatic fever, or impossible where muscle has disappeared as in myocardial infarction. Amelioration may be achieved if the muscle fibres are made to do more work under the influence of digitalis.

There may be hypertrophy of muscle to overcome a specific mechanical obstruction to cardiac output as in arterial hypertension or aortic stenosis or mitral stenosis; and when failure occurs it is muscle failure but related to the co-existing mechanical fault; and if the mechanical fault is removed then the heart muscle may once again prove adequate to the needs of the body. But the situation is rarely as simple as this. For with hypertension, for example, coronary artery disease may also occur, and this will cause ischæmia and death of heart muscle and then even a complete removal of the mechanical cause (as in coarctation of the aorta successfully overcome by surgery) will not wholly remove the heart failure. And with rheumatic valve disease there is commonly a coincidental damage to the heart muscle with inflammation and fibrous replacement, and then repair of the valve may still leave more or less muscle insufficiency. Heart muscle disease is more important than heart valve or mechanical disease, and where possible valve or mechanical disease should be cured while the heart muscle is still relatively normal. Coarctation of the aorta is an example; the heart muscle is normal and when the

mechanical cause of muscle hypertrophy has been removed early enough a normal life may be expected (remembering always the possible presence of associated anomalies such as intracranial aneurysm, bicuspid aortic valves, aortic cystic medianecrosis.

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$$\frac{4\cdot5\ (\text{art.}) + 10\ (\text{vein})}{2} = 7\cdot25 \text{ vols. per cent}$$

or Capillary blood may be more completely reduced

$$\frac{0\cdot5\ (\text{art.}) + 13\cdot5\ (\text{vein})}{2} = 7 \text{ vols. per cent}$$

Anæmia, by reducing the available quantity of Hb may make cyanosis impossible, e.g. 30 per cent Hb = less than 5 grams Hb. Hence absence of cyanosis with anæmia does not exclude anoxia.

Conversely, polycythæmia may make cyanosis obvious though its presence does not necessarily imply anoxia, e.g.

say Hb = 200 per cent Hb 30 grams

Arterial desaturation 1 vol. per cent

Venous desaturation 11 vols. per cent

$$\text{Capillary desaturation } \frac{1 + 11}{2} = \frac{12}{2} = 6 \text{ vols. per cent}$$

$$6 \times \tfrac{3}{4} = 4\tfrac{1}{2} \text{ grams Hb reduced}$$

so that in otherwise normal conditions a person who has *polycythæmia is nearly cyanosed* and only a minor change will cause cyanosis.

And because the blood in polycythæmia is thicker than normal and more viscous it flows slowly, more oxygen is extracted and there is more reduced Hb. The blood volume is also larger and the capillaries at all times fuller than normal, venous stagnation is usual, and with even slight body cooling cyanosis may be intense. Hence cyanosis with polycythæmia because it is peripheral does not imply anoxia. It is also thought that in polycythæmia the fact that blood is present in thicker layers is responsible for some of the dusky colour.

Cyanosis is of two varieties—three if we include states in which Hb is changed to Sulph- and Meth- hæmoglobin. The two forms which concern surgeons are:

(1) Central cyanosis.

(2) Peripheral cyanosis.

(1) Central Cyanosis

Central cyanosis occurs in two distinct ways:

(*a*) In one there is venous (5·5 vols. per cent unsaturated) blood in the systemic arteries (0·5 *vols. per cent unsaturated) owing to some* mechanical abnormality which permits this admixture. (The anatomical conditions in which this admixture may happen are considered when these specific abnormalities are discussed.) It can be shown that if a normal degree of desaturation of blood takes place in the capillaries (i.e. 5 vols. per cent) then arterial blood must be 4·5 vols. per cent unsaturated if cyanosis is to occur. By calculation

$$\frac{4\cdot5\ (\text{art.}) + 9\cdot5\ (\text{vein})}{2} = 14/2 = 7 \text{ vols}$$

$$7 \times \tfrac{3}{4} = 5\tfrac{1}{4} \text{ gram Hb reduced}$$

Pulmonary infection with heart disease is not a contraindication to surgery, rather to the contrary. Syphilis does not preclude operations upon the great vessels.

Acute pulmonary œdema is commonly fatal.

Angina pectoris is unpredictable. It may appear with worry and disappear when the worry goes. Likewise it is not uncommon at the menopause for a patient to suffer transitory angina pectoris.

The prognosis of heart disease with obesity is rather less bad because fat can be removed and improvement may result; on the other hand a gain in weight may adversely affect heart disease.

Subacute bacterial endocarditis as a complication is often not recognized and accounts for some bad surgical results.

Cyanosis

Everyone recognizes cyanosis when he sees it. Dilated capillaries make it easier to detect by increasing the quantity of reduced hæmoglobin in the tissues. Contracted capillaries make cyanosis inconspicuous. Increased CO_2 in the blood dilates the peripheral vessels and also causes Hb to become more easily reduced, and if anoxia occurs when CO_2 content of the blood is high, cyanosis is intense. Reduced CO_2 content in the blood constricts peripheral vessels and also causes Hb to resist reduction; if anoxia occurs with low CO_2 content, cyanosis will be inconspicuous.

When cyanosis is present at least 5 grams of Hb in each 100 ml. of blood are in a reduced state. If we consider the normal condition of the blood this statement will have more significance.

(1) Normal blood can take up 20 ml. of O_2 per 100 ml. when it contains 15 grams Hb per 100 ml This means that $\frac{3}{4}$ gram Hb takes up 1 ml. O_2.

(2) Normal arterial blood contains 19·5 ml. of O_2 per cent. This means that it is 0·5 ml of O_2 per 100 ml. unsaturated: this is usually expressed as 0·5 vols. per cent unsaturated.

(3) Normal mixed venous blood in the right heart contains 14 vols. per cent O_2, i.e. it is 5·5 vols. per cent unsaturated; it has given up 5 vols. per cent in the capillaries, which may be taken as an average O_2 loss.

(4) Capillary blood, halfway between arteriole and venule may be taken as being 3 vols. per cent unsaturated, i.e.

$$\frac{0{\cdot}5\ (\text{art.}) + 5{\cdot}5\ (\text{vein})}{2} = 6/2 = 3 \text{ vols. per cent}$$

From the foregoing, 3 vols. per cent unsaturated means $2\frac{1}{4}$ grams Hb reduced (1 ml. of O_2 is carried by $\frac{3}{4}$ gram Hb) and therefore 5 gram Hb reduced means 7 vols. per cent unsaturated.

i.e. $$7 \times \tfrac{3}{4} = 5\tfrac{1}{4} \text{ gm. Hb reduced.}$$

this degree of unsaturation in the capillaries will produce cyanosis. And this state (7 vols. per cent) of unsaturation in the capillaries may come about in either of these two ways, or in *one of a variety of intermediate changes:*

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$$7 \times \tfrac{3}{4} = 5\tfrac{1}{4} \text{ gram Hb reduced}$$

(*b*) In the other, venous blood is not adequately oxygenated in the lungs.

(2) Peripheral Cyanosis

In peripheral cyanosis the blood flows more slowly and gives up more O_2 and therefore carries more Hb in a reduced state. Calculation will show that cyanosis will appear when 13 vols. per cent (N = 5 vols. per cent) of O_2 are extracted in the capillaries

$$\frac{0{\cdot}5 + 13{\cdot}5}{2} = 14/2 = 7 \text{ vols. per cent unsaturated.}$$

$$7 \times \tfrac{3}{4} = 5\tfrac{1}{4} \text{ gram per cent reduced}$$

The cause of the peripheral cyanosis is any condition which causes capillary stasis. When the cyanosed part is found to be warm the cause may be taken as central; and in assessing the significance of cyanosis the colour of the tongue, which is warm and moist, is used as an indicator.

It will be clear that in many cases central and peripheral cyanosis will co-exist.

When there is doubt the matter can be settled at once by obtaining a sample of arterial blood for gas analysis.

When there is an interventricular septal defect and the aorta is so placed that blood emerging from the right ventricle enters it the desaturated blood mixes with the saturated blood from the left ventricle and central cyanosis results. With exercise venous return increases and the pressure and volume output of the right ventricle increases and, if there is pulmonary obstruction, a larger proportion of desaturated blood enters the aorta and central cyanosis increases till it becomes intense and effort can no longer be continued and syncope may occur from anoxia.

When a communication exists between the right and left sides of the heart, blood may be shunted through it in a direction away from the chamber with the higher pressure, e.g. in an uncomplicated A.S.D. blood flows from left to right, but when there is straining or crying the flow may be temporarily reversed with right to left shunt and cyanosis. In the later stages of an A.S.D. when the prolonged burden of the left to right shunt has caused pulmonary hypertension, right ventricular and right auricular hypertrophy, there may be a permanent reversal in the direction of the shunt with permanent cyanosis. This is specially the case when respiratory infection further increases the right heart pressures. Where the hiatus is large, simultaneous shunting in both directions occurs and is not rare.

A similar reversal of the direction of the shunt may occur when a patient ductus arteriosus is eventually complicated by pulmonary hypertension; the cyanosis which then occurs effects the lower half of the body; it is intense in the feet, with clubbing of the toes, and gradually disappears about the level of the waist.

Dyspnœa in Heart Disease

Dyspnœa occurs in heart disease but the causes are not identical in all cases In *constrictive pericarditis* dyspnœa occurs when the increased metabolic requirements of effort are not met by an increased cardiac output—the exact mechanism whereby this causes the sensation of dyspnœa is unknown, but it is presumed to originate in reflexes which originate centrally and peripherally. The lungs are normal and such patients can lie flat without discomfort.

In Fallot's tetralogy, with effort there is increased venous return and a larger than resting quantity of desaturated blood passes into the dextraposed aorta with increased cyanosis and a drop in the arterial oxygen saturation and to this the dyspnœa is attributed. Dyspnœa is severe and may be associated with a squatting posture; this is adopted by the patient to decrease respiratory discomfort, and various explanations have been offered as to how it does so. These patients can also lie flat without discomfort.

In isolated pulmonary stenosis dyspnœa occurs on effort and may be due to insufficient cardiac output to meet the needs of metabolism and to reflexes which originate in the right side of the heart with the increased pressure there caused by the venous return. The patient lies flat without discomfort.

In mitral valve disease the left auricular pressure is raised and there is impairment of the flow of blood through the lungs which contain a larger volume of blood than normal. The pulmonary capillary pressure is raised, the capillaries are dilated and the lung tissues are congested and œdematous. The lungs are less elastic and are stiffer to move. Effort increases the pulmonary plethora and exaggerates inelasticity. The respiratory muscles do more work than normal to produce ventilation, and this excess activity is registered as abnormal breathing. Reflexes arising within the lungs may also play a part in creating the sensation of dyspnœa.

In advanced cases not only are the lungs stiff but there is reduced vital capacity because there is more blood in the thorax, the heart is larger than normal, there are pleural effusions, the liver is enlarged and ascites limits diaphragmatic movements. Such patients are short of breath at rest, are unable to lie flat or even recline, but are least uncomfortable when sitting upright. Any effort, fear, or excitement which raises the heart rate will reduce the duration of diastole, reduce filling time of the left ventricle and cause greater overdistension of the lungs and so lead to acute pulmonary œdema. As soon as the tricuspid valve becomes incompetent in such cases there is considerable relief of dyspnœa at rest because the pulmonary plethora is relieved by the diminution of right ventricular output which results from the backflow.

In mitral valve disease the congestion and œdema of the lung tissues renders these structures more liable to recurrent infections which present the clinical picture of acute, subacute, or chronic bronchitis. There is with these infections a supra-added bronchial spasm with expiratory embarrassment and wheezing. These attacks increase the already present dyspnœa due to the mitral valve lesion, and the relief given by surgery consists in part in a reduction of these recurrent infections.

When there is hypertension or aortic valve disease or coronary artery disease which affects the left ventricular activity, in addition to effort dyspnœa (which may be considered as being in the main due to inadequate left ventricular action and consequent pulmonary congestion) there is another form of dyspnœa which occurs in paroxysms during the night. The patient is asleep and is suddenly woken with his chest tight, difficult breathing, often wheezing expiration, cough with little or no sputum, much sweating and either pallor or suffusion of the face. These attacks last a few minutes or sometimes as long as hours and though alarming are seldom fatal. During an attack the lungs are full of rales and rhonchi, there is triple heart rhythm and the blood pressure is raised.

The significance of these various causes of dyspnœa is that their varied mechanisms should be recognized and suitable steps taken to control the harmful effects before and at operation. In Fallot's tetralogy this can be done by decreasing the shunt through the

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Clubbing of the Fingers

Finger clubbing occurs with heart disease whenever there is cyanosis due to a shunt from the venous to the arterial circulations no matter what the specific abnormality may be which causes this shunting, and it often occurs with heart disease without cyanosis when there is bacterial endocarditis. It also occurs in suppurative and neoplastic disease of the lungs, and in some chronic infections elsewhere. It is occasionally found to be unilateral when an aortic aneurysm interferes with the circulation in one limb. The grosser degrees of finger clubbing in congenital heart disease with cyanosis are seen in those patients who have an associated arachnodactyly.

Finger clubbing (and toe clubbing which co-exists) is an hypertrophy of the soft tissue; the nails grow curved in their long axis, the finger-ends grow bulbous, the nail beds often are bluish-coloured and are "fluctuant" to touch. At times there is an associated overgrowth of the periosteum of the peripheral limb bones (hypertrophic osteo-arthropathy) and effusion into the joints and œdema of the peripheral tissues with pain and a sensation of heat. The blood flow in the fingers and limbs is increased, there is capillary dilatation, open A-V anastomoses and probably an increased lymph flow.

When the cause is removed the clubbing may disappear.

Air Embolism

There are three distinct ways in which air embolism may complicate operations upon the heart or the great vessels. In the past this accident has rarely been of such magnitude as to cause clinical symptoms; but when it occurred the patient generally died. As the surgery of the open heart becomes a practical possibility the question of air embolism assumed a new importance. The treatment is always prophylactic, and sometimes active. It is urgent.

(1) During inspiration the pressure in the cavæ and the innominate veins is low and if a small lateral tear be inadvertently made in one of these vessels air may be sucked into the venous system. When this happens the surgeon may or may not be aware of a hissing sound as air bubbles are sucked into the vein. If this accident should ever happen the traditional advice is to flood the wound with saline, occlude the vein and close the tear.

The actual mechanism by which such patients are killed is in some dispute but autopsy generally reveals a mass of frothy blood in the right ventricle and the assumption is that the air has acted as an "air lock" in a pump. If this is so then active treatment might save the patient. As soon as there is any indication that the heart has stopped beating the chest should be opened and cardiac massage, together with the other measures generally used in the management of cardiac arrest, should be vigorously applied. In this way it might be possible to re-establish the circulation. There is nothing to lose and everything to gain.

(2) In cardiac operations the heart is often approached across a pleural cavity and the lung may be adherent to the chest wall. Most pleural adhesions contain adventitious blood vessels and these form anastomotic connections between the systemic venous and the pulmonary circulations; many of the intercostal veins into which this blood passes flow into the azygos and so into the paravertebral systems of veins. When any part of this network is torn or opened in such a way that air can be sucked in, the bubbles can

aorta and increasing the flow through the pulmonary artery and lungs In Pick's disease by removal of the constricting pericardium and so increasing the cardiac filling and cardiac output. In mitral valve disease by making the valve more nearly normal and so decreasing pulmonary congestion with the increased stiffness of the tissues which it causes, and also by reducing the liability to infections with bronchial spasm. In aortic valve disease, coarctation of the aorta, and coronary artery disease by making the valve more normal, by excision of the coarctation, or by revascularization of the heart muscle to increase the left ventricular efficiency and reduce chronic pulmonary congestion and the risks of acute pulmonary œdema, paroxysmal nocturnal dyspnœa, and bronchial infection.

Drugs, such as digitalis which is essential in heart disease as it increases the myocardial force, morphia which reduces pain and dyspnœa and is specific in the treatment of paroxysmal nocturnal dyspnœa, correct position of the patient to increase respiratory capacity, removal of pleural effusions and œdema—all assist in bringing the suitable patient to definitive surgery at which the mechanical obstruction to cardiac action may be overcome.

Heart Disease and Pregnancy

When heart disease is present in a form which is amenable to surgical treatment, e.g. uncomplicated mitral stenosis, the problems when the patient is pregnant are relatively simple.

If there has never been and there is at present no heart failure, and the capacity for exercise is but moderately reduced, she will almost certainly go through pregnancy and parturition without difficulty, and surgery of the mitral valve should be delayed till after delivery.

But where there is great or increasing reduction of the capacity for effort, or where there has been in the past or is at present heart failure, surgery should be undertaken forthwith Medical control of heart failure is an essential preliminary to surgery.

The stage of pregnancy is immaterial (if possible avoiding the third month which is in any event the commonest time of miscarriage) and mitral valvotomy has been successfully performed even late in pregnancy.

Polycythæmia

When the oxygen saturation of the arterial blood is reduced below normal the bone marrow responds to this anoxia by increased production of red cells. As a result the total red corpuscle mass is increased and red cell counts of 10 millions and higher are found, but there is not a corresponding increase in the plasma volume and the blood is therefore thicker and more viscous than normal with hæmatocrit readings of 70 per cent and higher (normal 44–47 per cent).

As a result of increased blood viscosity the peripheral circulation is slower, with peripheral cyanosis, and *intravascular thrombosis may occur in particular in the brain, lungs,* and heart. In congenital heart disease with cyanosis there is therefore danger of intravascular thrombosis occurring when post-operative dehydration occurs This will be particularly so in hot climates and during the summer. In such conditions this danger applies in particular to infants and children who become rapidly dehydrated because of their higher metabolic rate.

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pass into the paravertebral system of veins and so up into the *venous sinuses in the skull* and cause symptoms by interrupting the cerebral circulation. Most cases of "pleural shock" are in this category.

This type of air embolism was not uncommon when artificial pneumothorax and cauterization of adhesions were routine methods of treating some cases of pulmonary tuberculosis. It can theoretically occur whenever pleural adhesions are divided. If the operation is being conducted under local anæsthesia the patient generally experiences a sudden catastrophe; he cries out, complains of chest pain and may or may not become unconscious. All grades of result, from mild and transient palsies to sudden death, can occur. There is no effective treatment.

(3) When the left side of the heart is opened air gets into the ventricle unless the opening happens to be at the top of the exposed parts. This accident causes death by coronary or cerebral air embolism: the bubbles can be seen obstructing the coronary vessels, and only urgent measures are likely to restore circulation to the myocardium. Geoghehan and Lam (1953) have shown the mechanism by which the air bubbles obstruct the coronary circulation. Whilst the proximal end of the column of air in the vessel moves onward with each ventricular contraction, the distal end remains stationary. The propulsive force is thus damped out by compression of the air. They were also able to show that some of the air could get through the capillaries into the coronary sinus. The basis of treatment is thus to raise the arterial pressure within the coronary arteries so that the bubbles may be pushed on into the coronary sinus. This can be achieved by clamping the aorta, massaging the heart vigorously, and aspirating residual air from the apex of the left ventricle. Whenever the left heart or the ascending aorta is opened special precautions must be taken to avoid air embolism.

Hypothermia

Definition. The term hypothermia is used to describe a state in which the body temperature is artificially lowered to a controlled level. It should not be confused with "hibernation," which is a natural state in the animal kingdom, although the two may be comparable in some respects. The words "freezing" and "refrigeration" are inaccurate.

Physiology. The object of hypothermia is so to lower the respiratory activity, and particularly the oxygen consumption, of the cells of the whole or a part of the body, that the circulation can be reduced or eliminated for much longer than usual. It has also been shown that the secondary changes due to enzymes, which are normally caused by hypoxia and which ultimately kill cells, can be delayed. The reaction of different tissues to cold varies greatly It has been shown for instance that entire limbs can survive complete anoxia for 50 hours at 2°C. It is believed to be safe to exclude the coronary or the cerebral circulation for about 10 minutes at 25°C. In applying hypothermia to a patient one must consider not only the effect upon the part under operation, but upon the organism as a whole. In the case of the myocardium the limiting factors are not completely known, but there is an important disturbance in potassium in the muscle and this is concerned in some way with an increasing liability to ventricular fibrillation as the temperature drops

The possibility of applying hypothermia to cardiac operations was first advocated by Bigelow and his colleagues (1950). It was shown that the total oxygen consumption of warm-blooded animals could be safely and reversibly reduced without incurring an oxygen

debt, so that the heart could be excluded from the circulation for periods of many minutes. It was further discovered that the heart could be opened under such circumstances, and hence that direct operations upon the chambers themselves were possible. The cooled heart continued to beat normally, even though the supply of blood has been cut off by clamping both vena cavæ. The heart rate was progressively diminished as the temperature fell. When the heart had been opened and the blood temporarily evacuated, the movements continued. Manipulations which involved the atria were better tolerated than those upon the ventricles, and a very cold heart was more susceptible to dangerous arrythmias than one at normal temperatures. Experimental animals can be safely cooled and rewarmed through temperatures of 38-20°C., and, when used in human beings, hypothermia has been restricted to this range. The idea that shock is increased by cooling does not hold, provided the "shivering phase" be eliminated.

Methods of Producing Hypothermia for Cardiac Operations. Whatever technique is used deep general anæsthesia is necessary to prevent shivering and other muscle reflexes. Without anæsthesia, shivering produces a high oxygen debt which defeats the objects of the method and can be lethal. The actual cooling can be done in a variety of ways of which the following have been used clinically. The first depends upon a combination of surface cooling and sympatheticolytic drugs. The second combines surface cooling and sedatives (as well as anæsthesia), and in the third cooling is achieved by using the blood stream. Each has its advocates and advantages; but it seems probable that some perfection of skin or blood stream techniques will ultimately be preferred.

The "surface methods" involve immersing the patient in cold water until the requisite drop of temperature has been achieved, or in the use of cooling blankets in which the patient can be wrapped. To cool the blood it is necessary to take blood out from an artery or a vein, to pass it through a refrigerating plant and then to reintroduce it into a vein. The apparatus required is simple and relies upon the patient's own heart to pump his blood through a small, cold, bypass. The actual choice of technique depends to some extent upon the size of the patient. Once cooled a patient remains at the depressed temperature until rewarmed.

Indications for Using Hypothermia. At the time of writing the technique provides one method of achieving a bloodless heart for sufficiently long to carry out certain intracardiac operations which cannot be accurately done in other ways. That is, it is especially suited to the closure of simple atrial septal defects and to resection of infundibular stenosis. In the future it may well be applied to the reconstruction or replacement of damaged valves.

The period of hypothermia should be as short as possible. For this reason the chest should be opened and the diagnosis confirmed before cooling is started. A wide exposure of the heart is essential. The risk of ventricular fibrillation must be realized, and the methods of control must be at hand. A completely dry heart is not necessary, and is seldom achieved because the coronary sinus remains open and aberrant veins may be present. Coronary air embolism is a serious risk whenever the left heart is opened. Rewarming should begin as soon as the heart has been closed.

In the future it is possible that hypothermia will be used in conjunction with other methods.

The Artificial Heart-Lung

The general trend of cardiac surgery is to achieve conditions which are such that the

heart can be opened and the necessary procedures carried out under direct vision. To this end it is necessary so to reduce the oxygen requirements of the myocardium and the brain that the flow of blood through the heart can be eliminated or reduced for a sufficient time, as for instance by the use of hypothermia; or the respiratory functions of the patient's heart and lungs must be taken over either by using some type of cross-circulation or an artificial heart-lung machine. It is possible that the final solution of these problems will involve a combination of the above methods, and already a great variety of partial short-circuits have been devised and applied.

The first surgeon to use such a machine successfully was Gibbon (1953) who repaired an atrial septal defect in an adult. The heart and lungs were excluded completely from the patient's circulation for 25 minutes and the operation was successful. He had spent many years perfecting the apparatus which had been tried out extensively on animals. Heart-lung machines are now in use in many clinics throughout the world.

The blood returning to the heart by way of the vena cavæ must be diverted into an apparatus which fulfils the following functions: It must be oxygenated and excess of carbon dioxide removed. To achieve this end it must flow through an atmosphere of oxygen and be spread out into such a thin film that all the cells come into contact with the gas. Clotting must be controlled by anticoagulants and although they can theoretically be neutralized it can cause bleeding difficulties in the patient after operation. The flow of blood through the apparatus and back into the patient must be accurately controlled both as to rate of flow and pressure. The passage of the blood through numerous external tubes and pumps can result in hæmolysis and other dangerous changes in the platelets: to reduce these risks the apparatus must be as simple as possible. A further problem is that, as the collection of sufficient blood from the great veins is difficult, the effective rate of perfusion is limited, and this can only be overcome by passing the blood rapidly through narrow tubes which increase the destruction of red cells. The tendency of the blood to froth and bubble must be eliminated, the pH and the temperature must be controlled.

It is apparent that such an apparatus will be expensive and its application requires expert medical and electronic knowledge. In its present form it is beyond the scope of routine surgery. The uses to which a heart-lung machine might be put are under trial. It can either be used to provide a complete or a partial by-pass of the heart. It has been tried in the management of various congenital cardiac deformities, in which correction could only be accurately achieved by operating upon the open heart under direct vision; and in patients suffering from mitral incompetence in whom the aperture of the valve ring was reduced in size by stitching. Some patients suffering from severe aortic stenosis who were too ill to stand an operation have been treated, using the apparatus to assist the pumping of the defective heart. It has also been suggested as having a place in the temporary management of some acute emergencies such as coronary thrombosis, cor pulmonale, pulmonary embolism and status asthmaticus.

Cross Circulation

Definition. The term cross circulation has been used to cover a variety of experiments devised to exchange some of the blood of one animal, or part of an animal, with that of another of the same species. The clinical application of these methods, which particularly concern cardiac surgeons is that they offer another way of obtaining a dry heart for

intracardiac operations. The principle involved is that whilst the heart of the patient is excluded from the duty of maintaining his own circulation, the blood is pumped to him by the heart and from the body of a suitable donor.

Physiology. In 1939 O'Shaughnessy demonstrated that the venæ cavæ of a dog could be clamped, so that no blood reached the heart, for 4 minutes and that most of the animals survived. Clinical experience had shown that patients who had sustained a serious hæmorrhage often survived with a low cardiac output for a long time. These observations led to the work of Andreason and Watson (1953) who occluded both the cavæ for long periods but left the azygos vein unobstructed so that the only blood returning to the heart came through the azygos system. Thus it was proved that a dog could survive with a flow of only 8–14 ml./kg./min. On flows of the azygos factor range Andreason says "the blood pressure sinks to 30–40 mm. Hg.; respiration is slow but regular; the heart contracts down, beats at about 40 per minute, but contractions are smart; pupils dilate and are sluggish. After 30 minutes re-establishment of the circulation is quite possible but requires care, as, *if the cavæ are released too quickly, the heart overfills and stops or* fibrillates." It was thus established that the animal could survive with a very much reduced coronary and cerebral circulation, but this alone was not sufficient to enable intracardiac surgery to be done. Andreason and Watson then showed that if the dog, whose heart had been deprived of filling blood by clamping both cavæ and azygos veins, was connected to another dog in such a way that some of the arterial blood of the donor was delivered to the aorta of the recipient through a small tube ending in the vicinity of the coronary arteries, the coronary and the cerebral flows were adequate to maintain life. It was of course necessary to extract from the recipient dog as much blood as it received and to return this to the donor. In these experiments the total volume of blood exchanged per minute was small and no complicated apparatus was required.

The Clinical Application of the Cross Circulation Principle. Lillehei (1954) was the first to operate upon patients using these methods. A donor of suitable blood group was found. The chest of the patient was opened widely by a trans-sternal incision and a plastic catheter was passed into, and along, the left subclavian artery so that its tip lay in the aorta. A second catheter was passed into the superior vena cava, and on into the inferior vena cava, across the right atrium. This catheter had two side openings, one at its tip and one which was sited in the superior vena cava. When the cavæ were occluded round this catheter all the blood which normally would have returned to the heart was diverted into the catheter. The arterial and venous catheters were then separately connected to a simple pump which assured that just as much blood was returned to the patient as was sucked out; the other ends of the two catheters were fixed beyond the pump into the femoral artery and vein of the donor. At the moment when it became necessary to exclude the heart of the patient, the circulation was as follows. Arterial blood from the donor was delivered to the aorta and hence to the coronary arteries of the recipient: this blood could not flow back in the chambers of the heart because the aortic valves were competent. The blood from the venous side of the circulation of the recipient was delivered to the femoral vein of the donor and the heart of the patient was empty from then onwards.

Using this technique Lillehei and his associates have operated upon a number of patients successfully. They have noted that the heart of the patient, though empty, continued to beat in a slow and regular fashion. Because the coronary flow was good

there were no arrhythmias, and as the heart was not actually pumping it could be displaced and manipulated without harmful effect. Ample time, that is up to 30 minutes, was available for intracardiac operation. These operations have been done at normal temperatures, and there has been no donor mortality.

Indications for Using a Cross Circulation. The indications have been limited and most of the patients have been children suffering from congenital heart diseases and the donors were adults. The following types of lesion have been successfully treated: ventricular septal defects, atrioventricularis communis, isolated infundibular stenosis and Fallot's tetralogy.

It will be noted that these lesions particularly involve abnormalities in the ventricles, and the reason for this is that as cross circulation do not involve hypothermia, ventricular fibrillation is not a dangerous risk. It would seem that cross circulation techniques may be preferred to hypothermia for operations upon the ventricles and other methods for closing atrial septal defects.

CARDIAC ARREST

Sooner or later every surgeon will be faced with the fact that the heart has stopped beating. Until recently this was accepted as a "fait accompli," but resuscitation is often possible, provided the diagnosis is not delayed and provided the essential manœuvres are immediately executed.

The heart stops beating in one of two ways. Either successive beats become less and less effective until movements cease, this is the usual type of arrest; or *ventricular tachycardia* develops and merges into *ventricular fibrillation*, and thence to asystole.

ASYSTOLE means that the heart has ceased to beat. It is the opinion of many that the type of asystole which occurs during the course of heart operations can usually be corrected; but it does not follow that the heart can be made to beat normally; and whilst the condition lasts the myocardium, the conducting tissue of the heart and the central nervous system are in mortal danger. In rare cases, where asystole has been overcome without myocardial damage, life cannot be restored because the respiratory centre is, in fact, dead.

VENTRICULAR TACHYCARDIA. In this condition the ventricles beat abnormally fast; the atria may or may not be affected. As this type of rhythm is likely to go on to ventricular fibrillation it should be regarded as an antemortem state and corrected without delay. It can generally be recognized by direct inspection and palpation of the heart during the course of a thoracic operation, but is likely to be missed in other types of surgery. In the normal course of events the heart shortens from apex to base and rotates transversely to and fro during each cardiac cycle. In ventricular tachycardia the beats are not only more rapid but they look different; the rotary movements are absent. The condition is dangerous in itself because the cardiac output is importantly diminished. In fact the heart is hardly pumping blood, in spite of its rapid contractions.

VENTRICULAR FIBRILLATION was first described more than 100 years ago and, until recently, was regarded as a fatal condition. All co-ordinated action of the myocardium has ceased, and the contractions of the ventricles are no longer controlled by the pacemakers. Ventricular fibrillation hardly ever reverts spontaneously to normal rhythm; but the heart continues to beat and to emit electrical impulses long after the patient is dead. As one looks at the ventricles the muscle fibres appear to be twitching and writhing;

these abnormal movements generally start near the region of the interventricular septum and spread out in all directions. The fundamental points to appreciate are that the heart is still alive, but it has ceased to pump blood; and that, provided the myocardium is reasonably normal, it can be made to pump effectively again.

Ventricular fibrillation cannot be diagnosed with certainty during the course of an ordinary operation unless the pericardium has been opened and the heart exposed to view. Palpation is not enough. It can be recognized if a continuous electrocardiographic record is available throughout the operation.

The underlying cause is hypoxia of the myocardium and this may occur in a variety of ways during an operation. There may be generalized anoxia due to inadequate ventilation of the lungs; the circulation may be temporarily obstructed during operations upon the heart itself; the coronary circulation may be impaired as a result of anæsthetic agents which cause peripheral vasodilatation; hæmorrhage may cause prolonged hypotension; certain drugs, such as quinidine, calcium and potassium salts, mercurial diuretics, chloroform, adrenaline, etc., predispose to the condition by making the heart irritable; coronary occlusions due to air embolism or pre-existing arterial disease are particularly dangerous, and hypothermia, beyond certain limits, is itself a cause.

The Diagnosis of Cardiac Arrest. From what has been said it is manifest that early and accurate diagnosis are essential if life is to be restored. If the chest is already open the surgeon will probably be aware of the emergency, but the first warnings are usually given by the anæsthetist in other types of operation. If the surgeon cannot palpate the pulse in a major vessel he must, without delay, expose or palpate the heart itself. The duty of the anæsthetist is equally urgent. he must ventilate the lungs with pure oxygen and this must be achieved within less than 2 minutes and maintained until an effective spontaneous beat has begun again. Coronary flow depends mainly on the level of the diastolic blood pressure because the vessels fill mainly during diastole.

A relatively small circulation is adequate to maintain life for a long time, and it is not certain that the supply of oxygen to the myocardium, the conducting tissue of the heart and to the brain, is the only requirement. A flow of fluid through these vessels is perhaps as important as a supply of oxygen and it may be that the removal of certain metabolites is important. The surgeon must proceed as follows as soon as he has diagnosed asystole or ventricular fibrillation.

The Treatment of Asystole.

CARDIAC MASSAGE must be done in such a way as to produce a circulation, and in practice this means a palpable pulse and a recordable blood pressure. This can only be done by holding the ventricles in the hand and squeezing them rhythmically at a rate of about 50–80 beats a minute. The emergency necessitates opening the pericardium in order to get the hand around the ventricles, and the best exposures are either through the linea alba and the diaphragm from below, or through the 5th left interspace in the thorax The former is the simpler in that no respiratory control is necessary if the patient survives· the second is more efficient both in diagnosis and treatment. The surgeon must choose the best for the individual case. To massage the heart properly considerable energy is necessary The procedure soon becomes tiring for the operator and reliefs must be available. Massage must be continued not only until the heart beats again, but until the myocardium regains tone, in practice this may be one or two hours.

Beck stresses the urgency and deprecates half-hearted measures: amongst other advice he stresses the following undesirable actions. Don't panic and don't waste time. Don't scrub anew, or wait for special instruments. Don't look for a stethoscope. Don't attempt to massage or squeeze the heart by pressing upon the chest wall or the abdomen. *Don't inject adrenaline or anything else through the chest wall.* Whatever else is done, cardiac massage and pulmonary ventilation are the forestays of treatment.

OXYGEN must be supplied continuously to the lungs by the anæsthetist who carries out artificial respiration using a closed anæsthetic circuit. This not only ventilates the blood but helps to pump it on through the lungs.

THE AORTA SHOULD BE HELD OR CLAMPED distal to the origin of the left subclavian artery. The object is to get whatever blood is flowing into the coronary vessels: for the brain cannot live if the heart dies.

INTRA-ARTERIAL TRANSFUSION may be beneficial. The simplest way of doing this is to take a transfusion set and run blood into the aorta as fast as it will go. The needle should be inserted above the point where the artery has been clamped in order to establish a retrograde flow to the coronary arteries.

THE SECOND STEP IS TO ESTABLISH NORMAL CARDIAC RHYTHM. Using the measures described above in many cases of cardiac asystole the heart will start to beat efficiently. But sometimes the heart responds to massage and other measures by passing into ventricular fibrillation, and, this is of no avail to the patient. Fibrillation must be stopped in all parts of the heart as soon as possible; adequate pacemakers must survive, and preferably one should dominate the others; the muscle must still be capable of contracting and this will have depended on the coronary blood flow during the period of asystole or fibrillation.

The Treatment of Ventricular Fibrillation. The only way of *immediately* abolishing ventricular fibrillation is to defibrillate the heart electrically. This involves the use of an apparatus which stimulates the heart without burning it, and the object is to pass a current through the heart which is sufficiently powerful to make all the muscle fibres refractory at the same moment. At St. Thomas's Hospital we have used an apparatus *designed to work from the mains of 220 volts at 50 cycles A.C.; it is held in constant* readiness during cardiac operations. The surgeon encloses the fibrillating heart between two spoon-shaped electrodes and applies one or more shocks, each of 1 second duration. This throws the heart into asystole, and massage is resumed until normal rhythm occurs. Several electrical shocks may be needed before the heart begins to beat efficiently. Using these techniques patients have been completely revived after periods of arrest of 60 minutes or more.

No mention has been made of injecting adrenaline or any other drug into the heart or the circulation. The reason is that whereas there is much to be said about the dangers of all drugs under these circumstances, there is nothing which can certainly be said in their favour. Adrenaline itself must be avoided because it predisposes to fibrillation, especially when the heart muscle is anoxic, or in the presence of chloroform or cyclopropane. The best advice a surgeon can have at the moment is not to inject any drugs into the heart; but if drugs are used noradrenalin is the safest way of maintaining peripheral vascular tone without affecting the heart adversely. If asystole persists a few cubic millilitres of 10 per cent calcium chloride may be injected into the right ventricular chamber with the object of making the inert heart muscle sensitive to external stimuli.

Controlled Cardiac Arrest. Kirklin, working at the Mayo clinic, has applied controlled cardiac arrest in patients connected to an extracorporeal circulation and requiring a cardiotomy. The heart is deliberately stopped by clamping the aorta and injecting a dilute solution of potassium into the aorta in the immediate vicinity of the aortic valve. The potassium percolates through the coronary arteries and the heart beat ceases immediately. At the conclusion of the intracardiac operation the aortic clamp is simply removed, fresh blood washes the potassium out of the coronary system and the heart beat returns spontaneously. This technique was first used experimentally by Melrose in England, and it is likely to become a routine procedure.

THE PREANÆSTHETIC AND POSTANÆSTHETIC PERIOD

It is neither the intention nor within the capacity of the authors to discuss here the available techniques of anæsthesia or the anæsthetic agents available for patients with heart disease. But certain points are made which have been found useful in dealing with these patients.

Premedication must be fully effective because of the risk of acute pulmonary œdema immediately before or during induction in patients who have tight mitral stenosis or a high pulmonary capillary pressure.

Postoperative Management. There is a tendency, possibly because the liver may be abnormal in long-standing heart disease with a raised central venous pressure, for the cardiac patient to remain anæsthetized for a longer time than another without heart disease under apparently similar conditions. Prolonged unconsciousness of this type may be confused with cerebral damage, due to clot or air embolism, or thrombosis in the carotid arteries resulting from occlusion to prevent embolism while manipulations are carried out in the heart, or to anoxia. In general the absence of unilateral physical signs at this time will be reassuring, e.g. the presence of both corneal reflexes and equal tonus on both sides.

Adequate analgesia is essential, for pain and fear may cause struggling and lead to pulmonary œdema. Morphia remains the drug of choice for the relief of postoperative pain, and heroin in suitable dosage is as effective and also has the advantage of being less liable to cause vomiting. The desire to experiment with the newer analgesics should be resisted; these drugs can be more safely established in less experimental fields of surgery before they are introduced into cardiac surgery.

Morphia is specific when there is pulmonary œdema. Full oxygenation is essential, and all fluids and air must be removed, or drained effectively, from the pleural cavities.

At regular intervals of time after the patient returns to the ward, both the radial pulses, and both the dorsalis pedis or posterior tibial arteries must be felt (it is useful to make certain that they are palpable before the operation!).

The fact that the pupils are equal should be verified when the pulses are felt, they are found to be irregular and unequal when cerebral embolism has occurred.

As soon as the patient wakens he should be asked to open his eyes and move his hands and feet. The ability to comprehend and to do this is more reassuring than the elicitation of many and various physical signs.

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OXYGEN must be supplied continuously to the lungs by the anæsthetist who carries out artificial respiration using a closed anæsthetic circuit. This not only ventilates the blood but helps to pump it on through the lungs.

THE AORTA SHOULD BE HELD OR CLAMPED distal to the origin of the left subclavian artery. The object is to get whatever blood is flowing into the coronary vessels: for the brain cannot live if the heart dies.

INTRA-ARTERIAL TRANSFUSION may be beneficial. The simplest way of doing this is to take a transfusion set and run blood into the aorta as fast as it will go. The needle should be inserted above the point where the artery has been clamped in order to establish a retrograde flow to the coronary arteries.

THE SECOND STEP IS TO ESTABLISH NORMAL CARDIAC RHYTHM. Using the measures described above in many cases of cardiac asystole the heart will start to beat efficiently. But sometimes the heart responds to massage and other measures by passing into ventricular fibrillation, and, this is of no avail to the patient. Fibrillation must be stopped in all parts of the heart as soon as possible; adequate pacemakers must survive, and preferably one should dominate the others; the muscle must still be capable of contracting and this will have depended on the coronary blood flow during the period of asystole or fibrillation.

The Treatment of Ventricular Fibrillation. The only way of *immediately* abolishing ventricular fibrillation is to defibrillate the heart electrically. This involves the use of an apparatus which stimulates the heart without burning it, and the object is to pass a current through the heart which is sufficiently powerful to make all the muscle fibres refractory at the same moment. At St. Thomas's Hospital we have used an apparatus designed to work from the mains of 220 volts at 50 cycles A.C.; it is held in constant readiness during cardiac operations. The surgeon encloses the fibrillating heart between two spoon-shaped electrodes and applies one or more shocks, each of 1 second duration. This throws the heart into asystole, and massage is resumed until normal rhythm occurs. Several electrical shocks may be needed before the heart begins to beat efficiently. Using these techniques patients have been completely revived after periods of arrest of 60 minutes or more.

No mention has been made of injecting adrenaline or any other drug into the heart or the circulation. The reason is that whereas there is much to be said about the dangers of all drugs under these circumstances, there is nothing which can certainly be said in their favour. Adrenaline itself must be avoided because it predisposes to fibrillation, especially when the heart muscle is anoxic, or in the presence of chloroform or cyclopropane. The best advice a surgeon can have at the moment is not to inject any drugs into the heart; but if drugs are used noradrenalin is the safest way of maintaining peripheral vascular tone without affecting the heart adversely. If asystole persists a few cubic millilitres of 10 per cent calcium chloride may be injected into the right ventricular chamber with the object of making the inert heart muscle sensitive to external stimuli.

Controlled Cardiac Arrest. Kirklin, working at the Mayo clinic, has applied controlled cardiac arrest in patients connected to an extracorporeal circulation and requiring a cardiotomy. The heart is deliberately stopped by clamping the aorta and injecting a dilute solution of potassium into the aorta in the immediate vicinity of the aortic valve. The potassium percolates through the coronary arteries and the heart beat ceases immediately. At the conclusion of the intracardiac operation the aortic clamp is simply removed, fresh blood washes the potassium out of the coronary system and the heart beat returns spontaneously. This technique was first used experimentally by Melrose in England, and it is likely to become a routine procedure.

THE PREANÆSTHETIC AND POSTANÆSTHETIC PERIOD

It is neither the intention nor within the capacity of the authors to discuss here the available techniques of anæsthesia or the anæsthetic agents available for patients with heart disease. But certain points are made which have been found useful in dealing with these patients.

Premedication must be fully effective because of the risk of acute pulmonary œdema immediately before or during induction in patients who have tight mitral stenosis or a high pulmonary capillary pressure.

Postoperative Management. There is a tendency, possibly because the liver may be abnormal in long-standing heart disease with a raised central venous pressure, for the cardiac patient to remain anæsthetized for a longer time than another without heart disease under apparently similar conditions. Prolonged unconsciousness of this type may be confused with cerebral damage, due to clot or air embolism, or thrombosis in the carotid arteries resulting from occlusion to prevent embolism while manipulations are carried out in the heart, or to anoxia. In general the absence of unilateral physical signs at this time will be reassuring, e.g. the presence of both corneal reflexes and equal tonus on both sides.

Adequate analgesia is essential, for pain and fear may cause struggling and lead to pulmonary œdema. Morphia remains the drug of choice for the relief of postoperative pain, and heroin in suitable dosage is as effective and also has the advantage of being less liable to cause vomiting. The desire to experiment with the newer analgesics should be resisted; these drugs can be more safely established in less experimental fields of surgery before they are introduced into cardiac surgery.

Morphia is specific when there is pulmonary œdema. Full oxygenation is essential, and all fluids and air must be removed, or drained effectively, from the pleural cavities.

At regular intervals of time after the patient returns to the ward, both the radial pulses, and both the dorsalis pedis or posterior tibial arteries must be felt (it is useful to make certain that they are palpable before the operation!).

The fact that the pupils are equal should be verified when the pulses are felt, they are found to be irregular and unequal when cerebral embolism has occurred.

As soon as the patient wakens he should be asked to open his eyes and move his hands and feet. The ability to comprehend and to do this is more reassuring than the elicitation of many and various physical signs.

THE SURGERY OF THE PERICARDIUM

ACUTE SUPPURATIVE PERICARDITIS

This condition is one in which there is pus in the pericardium. Since the introduction of antibiotics it has become rare; hence it may escape diagnosis and treatment.

Surgical Pathology. Suppurative pericarditis generally occurs as a part of some illness such as septicæmia or pyæmia, pneumonia or mediastinitis. It may be due to injuries and it can complicate hæmopericardium. It occurs when a subphrenic abscess or an infected hydatid cyst in the liver bursts through the central tendon of the diaphragm. It can complicate operations upon the lung, the heart, or the œsophagus. It does not occur as a solitary manifestation of inflammation, unless it follows some operation in the chest. The usual organisms are the staphylococcus, pneumococcus, and hæmolytic streptococcus.

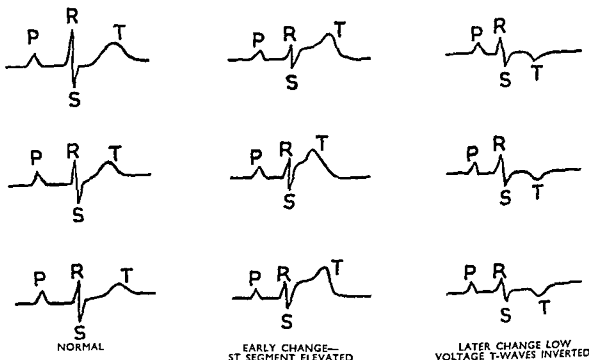


FIG 269. Acute suppurative pericarditis.

As a result of infection inside the pericardium the patient develops an effusion which is at first serous, but soon becomes purulent. How long this effusion takes to accumulate and how much fluid is poured out depends upon the organisms concerned; and the clinical signs and the progress of the illness are affected by these factors. If an effusion develops slowly the pericardium can accommodate a large volume without much venous congestion occurring. If it collects quickly as little as 100 ml. restricts the ventricles and atria and obstructs venous return by raising the intra-atrial filling pressures.

The fluid in the pericardium moves with alterations of position of the patient until it becomes localized by intrapericardial adhesions. When the patient *lies down* the exudate gravitates to the back of the pericardium and the front of the heart remains in contact with the front of the parietal pericardium. For this reason it is impossible to tell how much

fluid there is in the pericardium by examining the front of the chest; moreover a friction rub may be present anteriorly at a time when there is pus at the back and "muffled heart *sounds" will not be heard unless there is so much pus in the pericardium that the whole* sac has been distended. Aspiration through the front of the chest can result in a dry tap when the back of the pericardium contains pus.

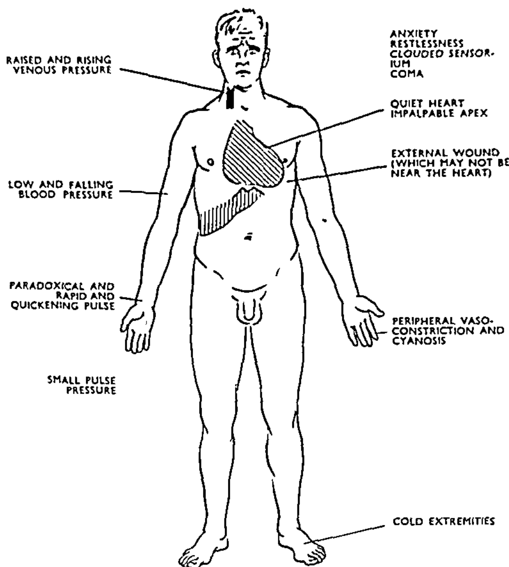


FIG. 270 Cardiac tamponade (see page 540)

As the *pericardium* becomes distended it assumes a *globular form*, as seen from the front, but the true shape of the sac is like two saddle bags astride the vertebral column. The backward bulge on the left side brings the sac into proximity with the left lower lobe bronchus, and the lower lobe may be atelectatic; in advanced cases the pus can be aspirated from the back of the chest, *but this is a bad manœuvre.*

Diagnosis. The diagnosis can be difficult because there are no signs pathognomonic of acute pericardial effusion and because the antibiotics effectively damp down the evidence of inflammation. The signs which may be present are pyrexia, tachycardia, and

those due to tamponade. The patient often complains of precordial pain and a pericardial friction rub may be heard in some cases. The pulse may be paradoxical and the heart quiet. The electrocardiogram shows low voltage curves in all leads; in the early stage the S.T. segments may be elevated, and later the T waves are often shallow and inverted. On screening, the shape of the heart tends to be globular and the pericardiophrenic angles may be obliterated. The normal movements of the borders of the heart are not seen. The lungs are clearer than normal.

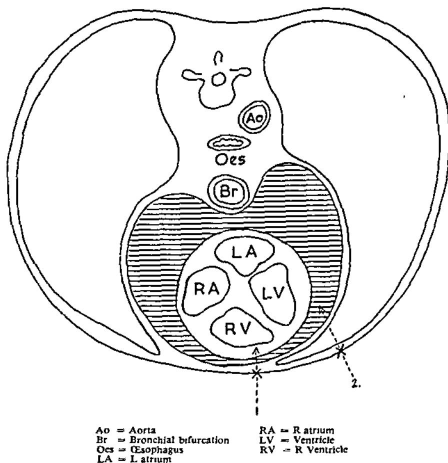


FIG 271. To illustrate the shape of the pericardium distended with fluid. (1) Costo-xiphisternal route of aspiration. (2) Trans-intercostal route of aspiration. Note that the needle traverses the pleural cavity.

The diagnosis rests upon *aspiration* and this should be done by passing the needle, between the xiphoid process and the left costal margin, upwards and backwards at an angle of 45 degrees and for a distance of about 10 centimetres. In this way the back part of the pericardial sac is explored, and, if the needle inadvertently enters the heart it does so at the apex which is the least vulnerable part. There is no need, and no justification, for aspirating in any other place if the tap is dry.

The differential diagnosis is from conditions which cause acute dilatation of the heart.

Treatment. It is justifiable to try repeated aspirations with instillation of the appropriate antibiotics; but this should only be continued as long as the pus can be effectively removed. As suppurative pericarditis is so often a part of some other inflammation it may be wise to temporize by aspirating until the other conditions have been controlled.

Surgical drainage will generally be needed if pus has formed, for antibiotics do not disperse pus.

Technique. Surgical drainage is done under local anæsthesia with the patient propped up on pillows. An incision 1½ in. long is made to the left of the linea alba and immediately below the costal margin. This incision is carried through the rectus sheath and the rectus abdominis muscle, until the transverse fibres of the transversus abdominis are seen. The surgeon keeps *superficial* to this muscle and, by blunt dissection, burrows up under the costal margin. In this way the pericardium can be reached from below without opening the peritoneum, incising the diaphragm or approaching the pleura. The diaphragm and the transversus abdominis are, in reality, parts of the same muscle; the fibres of each interdigitate behind the front of the ribs, and so by keeping superficial to the transversus abdominis one is also automatically above the diaphragm. The distended pericardium bulges downwards in a case of suppurative pericarditis and incision into it is not difficult. A corrugated rubber tube is inserted into the space behind the heart and the patient is nursed in the sitting position. Some surgeons advocate leaving a fine plastic catheter which reaches up from the superficial wound to the transverse sinus, so that irrigations and antibiotics may be applied locally. The pros and cons of such treatment are similar to those which cover irrigation of any serous cavity.

In this way, and in this way only, can dependent drainage be provided. The results of treatment on these lines are satisfactory, and Pick's disease is not likely to occur after convalescence.

CONSTRICTIVE PERICARDITIS

Definition. This disease has been called by many names including "Pick's disease"; but it was Richard Lower, Professor of Medicine at Oxford, who first described it in 1669 (Lower, R. (1669) *Tractatus de Corde*, London, p. 99), and Pick, in 1896, merely emphasized the accompanying cirrhosis of the liver. It is a condition in which the myocardium is at first relatively normal, but in which the visceral and parietal layers of the pericardium become involved in an inflammation which obliterates the sac and, ultimately, encases the heart in an unyielding bag of calcifying scar. The effects are to restrict cardiac filling and so to produce venous congestion and to diminish cardiac output.

Intrapericardial adhesions are often found in patients who have suffered no interference with cardiac function. The difference between these and constrictive pericarditis is that in the latter not only are both layers of the pericardium destroyed, but the sac is obliterated and the heart is strangled by scar and calcification.

Ætiology. The disease, as it occurs in Great Britain, is generally due to tuberculosis although the bacilli cannot be isolated at all stages of the illness. In a few rare instances it follows suppurative pericarditis or a traumatic hæmopericardium. It is not a complication of rheumatic fever. It is four times more common in men than in women and affects patients of all ages. Untreated it progresses slowly to a fatal termination once the signs of cardiac restriction have occurred.

Pathology. Constrictive pericarditis may be an end result of generalized tuberculous polyserositis. More often it is due to tuberculous mediastinal adenitis, and, in children this adenitis occurs as a sequel to a primary tuberculous complex in the lung. The enlarged nodes in these cases generally resolve without clinical caseation; but *sometimes* a caseating gland becomes adherent to the pericardium, in the vicinity of the *main carina*

or the left stem bronchus, and causes a lymphocytic effusion. This is similar in pathology to a serous pleural effusion and may follow the same course; in many cases it resolves, either spontaneously or as a result of appropriate medical treatment; but in some it becomes caseous and in a few autopsy specimens caseating glands have been shown to have ruptured into the pericardium. Once tuberculous pus has appeared in the pericardium the condition progresses to constrictive pericarditis in most cases. The pericardium itself becomes opaque, thick, and fibrous: it adheres to the adjacent pleura and limits the excursions of the diaphragm. The harmful effects are generally limited to those parts of the heart where the pericardium exists as a sac, and the inflammation does not involve the base so much as the apex of the heart.

The parietal pericardium may become more than an inch thick, and, in due season, the fibrous scar may be wholly or partly calcified. Between the two layers the pus inspissates and is granular, gritty, and semi-solid. In many patients the tuberculous process is limited to the pericardium from start to finish, but sometimes it involves the myocardium not only in replacement fibrosis, but also cold abscesses may form in the walls of the ventricles. These abscesses, upon rare occasion, can rupture into one of the chambers of the heart.

Tuberculous pericarditis is not necessarily rapidly or evenly progressive: there may be periods of apparent quiescence and others in which the symptoms and signs advance rapidly. In most cases the disease, untreated by surgery, eventually kills the patient. The effects of this type of pericardial destruction involve not only the heart which cannot fill properly and cannot vary its output, but indirectly the circulation of the blood; and it is the altered hæmodynamics which cause the most prominent symptoms. The patient suffers from slowly progressive venous obstruction of the superior and inferior vena cava which is manifest as raised venous pressure, enlargement of the liver, ascites, pleural effusions, and œdema of the legs and trunk. Cardiac cirrhosis commonly occurs, and some consider that a complete surgical cure is only possible as long as the cirrhosis is reversible, but it is likely that this opinion is based on the observation of those very rare cases where multilobular cirrhosis co-exists and causes ascites independently of the cardiac condition. A liver biopsy is helpful in some cases.

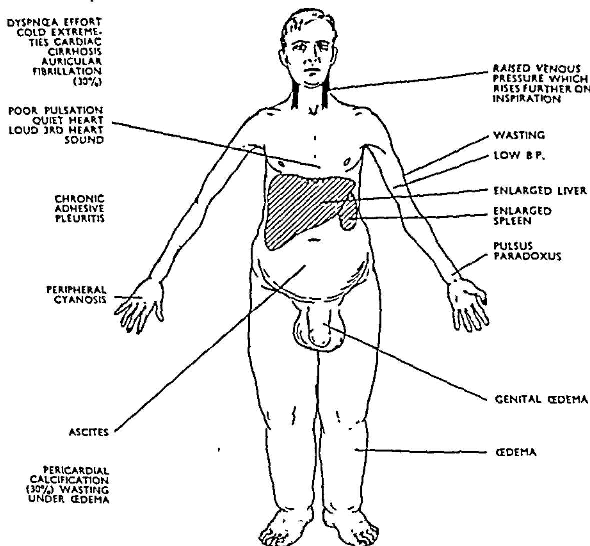
Surgical Anatomy. The tuberculous inflammation does not affect all parts of the pericardial sac evenly When a patient develops this disease he generally feels ill and goes to bed; lying in the recumbent position the pus in the pericardial sac gravitates to the back, and lower part. Thus the ultimate deformity of the dense scar occurs chiefly at the back and lower part of the heart, that is, over the left ventricle; the right ventricle is restricted to a lesser extent. In most cases the atria are not severely affected although the scar may form a strong annulus around the atrio-ventricular grooves. Such a band of scar can constrict the mitral valve from without in such a way as to produce the signs and symptoms of mitral obstruction.

It used to be said that the orifices of the venæ cava, as they enter the right atrium, were stenosed by scar: this is not so and, at operation, no attempt need be made to free these surgically dangerous areas.

The two layers of the pericardium may remain separated by inspissated effusion in which case the parietal layer is always much thicker than the visceral: indeed the visceral layer may be no thicker than a sheet of paper when the parietal is heavily calcified. The cardinal point for the surgeon to appreciate is that it is never enough to remove only

the parietal layer: a thin sheet of abnormal visceral pericardium restricts the heart as seriously as the massive and calcified carapace which is outside it. In many cases the two layers of the pericardium are inseparably fused, and quite often the scar tissue extends into the myocardium.

The coronary arteries on the surface of the heart are buried in the scar and it may be difficult or impossible to locate the descending branches. If these cannot be seen the



PROGNOSIS Prolonged disability with circulatory failure Commonest misdiagnosis—cirrhosis of liver Operative risks—15–20 per cent mortality Result of operation—excellent

FIG 272 Constrictive pericarditis

surgeon, who must preserve them at all costs, must rely on his knowledge of the normal anatomy, and leave the danger areas imperfectly decorticated.

Diagnosis. The onset of the signs and symptoms is usually gradual and occurs most commonly in the third or the fourth decade The patient is seldom pyrexial unless the onset is characterized by massive pericardial effusion. Many of the physical signs are like those of progressive congestive cardiac failure but the emphasis is somewhat different. The earliest manifestations are tiredness, dyspnœa on exercise, and swelling of the abdomen due to ascites. The legs usually become œdematous after the patient has noticed the distended abdomen due to ascites, pleural effusions may restrict respiration.

All patients suffering from restrictive pericarditis have a high venous pressure and venous pulsation is always present but may be hard to detect. In a severe case the liver is enlarged and may be hard and pulsating. Cyanosis is unusual, and when present is peripheral cyanosis.

The pulse is small, fast, and is often paradoxical; the systemic pulse pressure is low, and about one-third of all patients suffer from auricular fibrillation. The cardiac signs which differentiate restrictive pericarditis from heart failure of cardiac origin are these: in Pick's disease the apex beat is often impalpable and the heart is small; the heart sounds are difficult to hear and there are no murmurs, unless the mitral ring is constricted.

Important evidence is obtained by screening and radiography. There will be a "quiet" heart of normal size, and sometimes calcification which is always more obvious at the back and at the apex of the heart in the lateral positions, and, perhaps, some prominence of the superior and inferior vena caval shadow. Pleural effusions will also be obvious at this time.

Electrocardiography shows a low voltage curve, and the T wave is inverted or low, the P wave may be broad or notched. Cardiac catheterization is valuable in difficult cases but angiocardiography is seldom necessary.

The circulation time is increased and the cardiac output is diminished at rest and on exercise.

All other laboratory tests are within normal limits; but in advanced cases a liver biopsy may be done to assess the degree of cirrhosis The authors have personal experience of children whose liver biopsy showed gross cardiac cirrhosis and who have made a complete clinical recovery.

Treatment. It is now generally agreed that as soon as restrictive pericarditis has been diagnosed, and, long before calcification can be demonstrated in radiographs, active treatment should be instituted. In the early or "acute" stage of the condition the patient will have a serous pericardial effusion causing tamponnade: this effusion should be aspirated as necessary and streptomycin should be exhibited systemically and intrapericardially. Under such a regime the condition may resolve leaving no physical signs referable to the heart. If, in spite of medical treatment, aspirations become impossible, tuberculous pus accumulates and the signs of restriction persist, the sooner surgical treatment is instituted the better.

Some doubt has been expressed as to the wisdom of operating at a time when one may expect to find active tuberculous granulation tissue in the pericardium. It was felt that the disease might be spread to one or both pleural cavities or disseminated by way of the blood stream Those who adopted this policy were committed to wait for a long time and experience has shown that not only is this unwise because the heart itself may be seriously damaged; but that the tuberculous process is not spread by surgical intervention. The moment to operate is as soon as medical treatment of the pericardium begins to fail, but if the patient has active pulmonary tuberculosis this must be controlled first.

Shortly before operation it may be wise to diminish ascites or pleural effusions, by suitable diuretic drugs or aspirations; such effusions can impede respiration during induction of anæsthesia. But rapid or total removal of ascitic fluid can upset the protein balance of the body.

Operation. The object of operating is to free both ventricles by stripping off the viseral and the pericardial layers of the pericard um. It may be difficult, or impossible,

to do this thoroughly in the vicinity of the caval orifices, the auriculo-ventricular groove, or the coronary arteries. Fortunately the caval orifices are rarely obstructed and atria are not vitally important to pumping the blood.

The actual technique of access will vary according to the beliefs of individual surgeons. Those who follow the teaching of Holman, and who believe that both ventricles must be *completely* freed will prefer a trans-sternal approach opening both pleural cavities. If the main object is to free the back of the left ventricle where the scar, and calcification, is generally most dense, a left-sided, transpleural, thoracotomy gives excellent exposure. Many British surgeons feel that the most damaging part of the pericardial scar lies at the back of the left ventricle.

When the mediastinum has been reached, and in many cases numerous pleural adhesions will need division, the immobile, fibrous, fatty, and calcified tissue which encases the heart is seen. The phrenic nerve should be spared if possible, but, apart from this, the whole of the visceral and the parietal layers of the abnormal pericardium must first be cut and separated away from the left ventricle, and, after this, from the right ventricle if it is restricted. If the ventricles have been successfully freed they will enlarge visibly and not only will the active movements return but the heart will enlarge in diastole. Partial removal of the pericardium may satisfy the surgeon unaccustomed to this work but may not cure the patient. In some areas the visceral pericardium may be so densely adherent and the heart wall so thin from disuse or fibrosis that the ventricle is opened. To stop the hæmorrhage the surgeon should press on the spot with a finger and close the tear with a pericardial flap or mattress sutures. When the stripping has been completed antibiotic drugs should be left in the pleura, the chest should be closed in the usual way, and the pleural cavity drained for 24 hours. It will be found that quite a lot of sero-sanguineous discharge comes from the pleural drain during the first 24 hours, and adequate transfusions must be given both during and after operation.

Results. Excellent or satisfactory results may be expected in about 75 per cent of patients and the mortality of the operation has recently been reduced to between 10 per cent and 15 per cent. Deaths are due to bleeding or cardiac arrest at operation, sepsis, or reactivation of tuberculosis in other parts of the body. The mortality in patients over the age of 60 is high.

In successful cases the œdema and ascites disappear even though the venous pressure may not always be restored to normal, and the patients can return to a full and active life. In some the symptoms are not changed for several months and then, quite dramatically, the picture changes for the better. The author has seen complete recovery begin 6 months after an apparently unsuccessful operation, and has been forced to re-operate upon three patients whose symptoms persisted: these patients have all done well eventually. Cirrhosis of the liver is a serious complication but does not preclude a good result, because the liver may return to normal function after operation.

CARDIAC TRAUMA

Interest in this topic dates from the publication of Georg Fischer's monograph in 1868: he reported 452 patients wounded in the heart or the great vessels. He showed that heart wounds were not necessarily fatal and that sound healing of a wound could occur. Callender was the first Englishman to withdraw a needle from the heart and this he did at St. Bartholomew's Hospital in 1873. Many isolated operations had been performed

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Operation. The object of operating is to free both ventricles by stripping off the viseral and the pericardial layers of the pericard um It may be difficult, or impossible,

effectively. The systemic pressure falls due to blood loss or to shock, the intrapericardial pressure rises due to hæmopericardium and both these factors tend to limit bleeding.

Acute cardiac tamponade is the most characteristic immediate effect in those patients who survive the injury. It occurs as soon as a small amount of blood is confined within the pericardium. The blood may come from one of the chambers or from an injury to a coronary vessel. Apart from penetrating wounds the coronary vessels may be accidentally pricked when a pericardial effusion is aspirated, or the short sharp ends of the wire stitches, used after pneumonectomy to close the bronchus, can puncture the pericardium and cause bleeding.

Diagnosis. The diagnosis of a heart wound is often made upon the probability that the injury incurred is likely to have penetrated the mediastinum, but the skin wound is not always near the heart. The signs and symptoms vary. When the wound is small and little bleeding has occurred, as sometimes happens after a needle has been thrust into the myocardium, there may be no symptoms. At the other extreme is the patient who has lost a lot of blood, who is suffering from irreversible circulatory failure, and respiratory embarrassment due to hæmothorax. Sometimes there is a small wound on the front of the chest and from this spurts of bright blood emerge with each heart beat. If one of the coronary vessels has been severed the patient may have the signs of myocardial infarction. The most favourable cases are those who have *cardiac tamponade*.

The patient generally has a small penetrating wound, and blood may be trickling from this or emerging in small spurts. There is shock, which is often out of proportion to the amount of blood which seems to have been lost; the heart sounds are faint and the pulse is feeble and paradoxical; the arterial pressure is low and the venous pressure is high. The patient is dyspnœic and restless. A radiograph taken in the early stages may show no deviation from normal, but screening if it is possible reveals a decrease in the pulsation of the heart shadow. The diagnosis is seldom certain upon clinical signs and may depend upon aspiration.

Treatment. Restlessness and shock are treated with morphia, oxygen, and transfusion. The superficial wound is dressed and a radiograph of the chest is made. If the circulation deteriorates pericardial aspiration, by the subcostal route, is done immediately and the condition of the patient can often be dramatically relieved by taking off as little as 20 ml. of blood. Aspiration can be repeated with advantage on one or two occasions. But if the surgeon believes that bleeding is still progressing, open operation is urgent and imperative. Little, if any, anæsthetic is necessary in the early part of the operation. An anterior thoracotomy is generally made upon the side of the wound. On opening the mediastinum the pericardium can be seen distended with partially clotted blood. As soon as the pericardium has been incised tarry blood is extruded, and, if the patient is still alive, the heart fills and beats vigorously and consciousness returns so that anæsthesia becomes necessary. When the tamponade has been relieved bleeding from the heart may start again and must be controlled by digital pressure and sutures. On closing the chest the pericardium and the mediastinum should be temporarily drained into the pleural cavity.

Results. The patient who has a large hole in the heart or the great vessels does not reach a surgeon; but it should be possible to save those who reach the hospital alive. Treatable heart wounds are not always war emergencies, they occur also in civil practice and the mortality has been reduced to 10 per cent in some clinics. It is agreed that

upon the heart before the end of the century, and interest was further stimulated during the two world wars and culminated in Harken's work. The latter removed over 100 foreign bodies from the heart and great vessels without a death.

Many different forms of cardiac trauma are now recognized, and the problem no longer comprises the question of "penetrating wounds" alone. Only those aspects which concern the surgeon will be described in this chapter, although others such as the effects of a cold, heat, drugs, and electric currents upon the heart will certainly be important to operations in the future.

Closed Cardiac Injuries

The possibility that heart wounds could be caused by non-penetrating trauma was first envisaged by Oluff Borch of Copenhagen in 1676. Recently Warburg has written a book on this topic. His chief conclusions were these. Forensic material proves that closed cardiac injuries are relatively common, and the records of published cases show that all are not fatal. Tears and perforations of the myocardium may not be immediately fatal. The injuries which can occur include partial ruptures of the heart or the great vessels, avulsion of chordæ tendinæ (producing traumatic incompetence) and disruption of valves. They are more common in civil than in military practice, and can be caused by thoracic or abdominal blows and crushes. The trauma is usually violent and is often over the heart, but the ribs may be intact. The usual signs and symptoms are pain and evidence of tamponade in the acute phase: in those who develop symptoms slowly there can be a latent period of several weeks' duration. The late manifestations are due to recurrent pericardial effusions and disturbances of rhythm or coronary thromboses. The commonest late cause of death in those who survive the accident is congestive heart failure.

The surgeon can intervene with hope of success in several of these conditions. Tamponade can be relieved (q.v.); injuries to the great vessels have been sutured; traumatic aneurysms of the heart have been successfully excised, and it is possible that wounded valves might be repaired or replaced.

Penetrating Wounds

In many parts of the world, heart wounds due to knives, icepicks, stilettos and daggers are common; and suicides often try to push some object into the heart.

Surgical Pathology. Not all wounds are obvious, immediately fatal, or easily diagnosed. A penetration which merely opens the pericardium is of no significance; a small effusion may collect but seldom requires aspiration or other treatment: the hole seals off spontaneously. A long rent in the pericardium can be dangerous if it allows a cardiac hernia

A large wound of the heart is immediately fatal; it produces exsanguination into the *mediastinum, the pericardium and the pleural cavity. The pleural cavity fills* with blood, and, as the capacity of one pleural space in an adult exceeds 7 pints, the patient can die without external bleeding.

Statistics show that the order of frequency with which the various parts are wounded in those who survive is the right ventricle, the left ventricle, the aorta, and the pulmonary artery, and the atria. If the wound is not too large there are several factors which help to staunch the flow of blood. Wounds involving the atria are more dangerous than those in the ventricles: in the latter the thick interlacing muscle bundles often shut off incisions

An object carried along in a vein into the heart may impact in the atrium, be held in the tricuspid valve, or become entangled in the chordæ tendinæ. It may lodge in the auricular appendage. If it negotiates these obstacles it will come to rest in a branch of the pulmonary artery in the lung. Such an object, seen in a radiograph, looks as if it is lying in the interstitial tissue of the lung, and unless the surgeon is wise to this possibility, it will never cross his mind that the foreign body is inside a branch of the pulmonary artery, and massive hæmorrhage may occur if he tries to take it out without this knowledge. Difficult corners and sharp bends in blood vessels can be negotiated in a way which theoretically may seem impossible. A foreign body temporarily lodged in the right atrium may fall back into the caval system; an object which enters the jugular vein can drop through the atrium and lodge in a pelvic vein. In this way a foreign body can sometimes be moved from one place to another by positioning the patient.

Foreign bodies in the chambers of the heart, whether they have got there through the chest wall or by migration, can remain mobile and free for at least 8 weeks; but some become fixed within a few hours. Small radio-opaque objects have been seen, on screening, to be bouncing about inside one of the heart chambers like a ping-pong ball on the apex of a jet of water. Once inside the heart a foreign body generally becomes enmeshed in fibrin and shut off from the swirl of the blood like a chrysalis inside a cocoon. The fibrin organizes to form a capsule and such an encysted tumour may remain harmless for years; on the other hand it may not.

Sharp objects such as hairpins and toothpicks can perforate the heart, and perforation can occur without suppuration; they generally cause fatal hæmorrhage in the end. Partial perforation is a cause of recurrent pericardial effusion.

Signs and Symptoms. Migration from a peripheral vein to the heart or to the lung is symptomless unless the object is infected or unless it perforates the vessel. Even if it gets swept out of the ventricle into the pulmonary artery, so that it impacts in the lung, embolism and infarction do not occur. Perforation and infection are the dangers on the right side of the heart.

Migration of a foreign body out of the left ventricle into a systemic artery differs importantly from the silent movement in the venous side of the circulation because ischæmia of the brain or gangrene of a limb are likely to result. There is no way of predicting which foreign body in the heart will stay there and which will be shot off into the systemic circulation. Indeed the onset of signs and symptoms is generally as sudden as it may be unexpected. A foreign body impacted in the heart itself can cause a variety of upsets, including arrythmias, pain, dyspnœa, sudden death. It may lead to a cardiac aneurysm, embolism, abscess, thrombosis, progressive myocardial fibrosis, and hæmorrhage from perforation. Those held in the pericardium or the myocardium are particularly likely to cause repeated pericardial effusions. Large objects are more dangerous than small, rough things are worse than smooth and sharp objects do more damage than blunt ones.

The signs and symptoms may date from the day the patient was injured; but the foreign body may lie dormant throughout a long life. In every case the probability that complications will eventually develop and threaten life should dominate management of the problem.

Foreign bodies inside the cardiovascular system may carry organisms in with them. They may then continue to be symptomless for some time or they may give rise to

temponade should at first be treated conservatively, because the mortality under open operation is still high: this is partly due to technical mistakes and partly to the fact that some cases are hopeless from the start. Success depends upon doing a sound operation; there is no place for hurried and rough manipulations and the surgeon who has not considered these problems and is suddenly faced with the emergency of treating a heart wound will be lucky if his patient lives to tell the tale. Beck has stressed that most of the patients are young people with strong, normal hearts. If you do the proper thing they have a tremendous "come back" and no postoperative disability.

FOREIGN BODIES IN THE CARDIOVASCULAR SYSTEM

A variety of foreign bodies have been found in the cardiovascular system and the accidents have occurred in civil as well as military practice. The cases are seldom accurately diagnosed in the first instance.

Portal of Entry

Into the Veins

A foreign body can get into a vein without the patient having any knowledge of its presence. The commonest portal of entry is through the liver and thence into the hepatic vein and the inferior vena cava. A small object such as a fragment of cardiac catheter or a hypodermic needle can break off and be carried away to the heart. Perforation into a vein can occur when a radium seed or needle lies adjacent to it; and objects have entered the venous system from the alimentary canal, or by penetrating the pyriform fossa into the jugular vein, or from the duodenum into the inferior vena cava.

Into the Heart

Foreign bodies generally get into the heart by passing through the wall of the thorax, or by venous embolism, and they may lodge in the pericardium, myocardium or in one of the chambers. A foreign body in the left side of the heart gets there as a result of a penetrating wound of the heart or of one of the pulmonary veins: by perforation of the *lower œsophagus into the back of the left ventricle or into the left atrium*: by paradoxical embolism through an interatrial or interventricular septal defect.

Into a Systemic Artery

Apart from the aorta, into which small objects may enter directly, foreign bodies get into the arterial system by embolism from the left side of the heart.

Surgical Anatomy and Pathology. In all parts of the cardiovascular system the wound of entry heals quickly unless contaminated, and a short time after penetration it cannot be assessed exactly how a foreign body has got into the circulation. An aneurysm seldom occurs at the point of entry unless two adjacent vessels artery and vein are injured.

Once inside a vein a sterile foreign body is influenced by forces which are often opposed. The blood stream tends to sweep it towards the heart: gravity can pull it in the other direction. Objects which float rise upwards; heavy things fall down; hence migration and retrograde migration Movement of a foreign body may occur at any moment; it cannot be anticipated, or prevented except by removing the object from the vessel in which it lies.

Diagnosis. The site of the tumour determines the clinical picture produced in each case. A myxoma of the left auricle simulates mitral stenosis and leads to pulmonary hypertension, cardiac failure, and systemic emboli; the latter may be of clot or tumour fragments. Murmurs are variable or absent, and this point may suggest that the case is not one of mitral stenosis. A myxoma of the right auricle imitates vena caval obstruction without cardiac murmurs or arrhythmias. Relentless progress of cardiac failure, despite adequate obvious cause and despite rest and digitalization, is one of the diagnostic criteria. An abnormal outline on radiography has also been described. Innocent tumours do not interfere with the conduction mechanism, and electrocardiograms are normal. A myxoma situated in the right atrium may be outlined by dye injected into a peripheral vein, or by angiocardiography using a catheter. In the left atrium the diagnosis has most often been made at operation for supposed mitral stenosis. In such cases the mitral valve is normal to palpate and there should be no difficulty in distinguishing the features of a myxoma.

Sarcoma. Primary malignant growths of the myocardium have been recorded; they occur particularly in children and are sarcomatous. They differ clinically from myxomata because of the speed of growth and they cause hæmorrhagic pericardial effusions.

Treatment. If a myxoma has been diagnosed it can be most safely removed by cooling the patient, clamping off the venous return to the right atrium, as well as the aorta and the pulmonary artery, and then deliberately opening the right atrium and cutting the mass away from its point of attachment. If the tumour is in the left atrium it is probably wise to approach it from the right side across the septum. Particular care must be taken not to fragment the tumour which is soft and friable, and not cause air embolism.

Bailey has described a method of removing a myxoma from the atrium. If the tumour is in the left atrium he advises that the finger be kept in the valve whilst the tumour is being manipulated. Insert a purse string stitch in the wall of the atrium around the site of the proposed incision. The latter will vary in size according to the size of the tumour. Incise the atrium, having temporarily put an arterial clamp across the aorta and the pulmonary artery. The object of this clamp is to prevent air embolism into the coronary or the systemic circulations whilst the heart is open, and to reduce the return of blood to the left atrium from the pulmonary vascular bed: the clamp can be kept in place for about 2 minutes. Having incised the atrium the tumour can be expelled by the finger which is still in the auricular appendage, and the bleeding controlled by tightening the purse string stitch immediately the tumour is out. The principal danger of this manœuvre is air embolism. In the future it is probable that all problems of this kind will be solved by using an extra-corporeal circulation.

Secondary Cardiac Tumours occur more often in the right side of the heart and are found in about 3 per cent of patients dead of malignant disease. The majority occur in the wall of the ventricle and are not amenable to treatment. Their importance to surgeons is that a quarter cause a bloodstained pericardial effusion.

Hydatid Disease. Hydatid cysts sometimes occur in the heart or the pericardium: many have been reported in the literature. The majority of the patients die because the parasite ruptures into one of the cardiac chambers and disseminates the disease or causes fatal bleeding; some patients have survived this accident and have died of multiple secondary *echinococcosis* effecting either the lungs or the systemic viscera. Degeneration, calcification, and infection have been noted in these cysts.

pyrexia of unknown origin. Pyæmia, septicæmia or abscess can occur, and embolic abscesses will not be at the same place as the foreign body which causes them.

Treatment. The guiding principle is that all foreign bodies should be taken out because they are potentially dangerous: the exception is when the surgeon estimates the risk of operation as higher than the probability of serious complications.

It may be difficult or impossible without surgical exploration to say whether a foreign body lies in the cardiovascular system or not, but any object lodged in a blood vessel or the heart is a potential danger to life and wherever it is it should be taken out whenever possible. In practice this resolves itself into the treatment of foreign bodies lodged in the heart, in a branch of one of the pulmonary arteries, or suddenly impacted in a peripheral vessel producing ischæmia of the limb. If there is no special contraindication a foreign body must be taken out: but argument ranges about the proper treatment of an object lodged in the heart itself. The technical details of the operations which may have to be performed have been so perfected that the onus is now upon the surgeon to say why a particular object should not be taken out of the heart rather than to say why an operation should be advised.

There are *three* clinical phases in the history of these patients in which a decision must be taken.

(1) In an emergency the concern is to save life and the presence of a foreign body in the heart is of secondary importance.

(2) During the period of convalescence the decision to operate or not depends upon the belief that the foreign body constitutes an immediate danger to life and health, or that late complications can be avoided or that limitations of cardiac function can be improved without exposing the patient to mortal hazard.

(3) When a long-retained foreign body has caused cardiac arrythmias or serious neurosis because of the patient's knowledge of its presence, removal may be advisable. Sauerbruch, writing after the first world war, emphasized that retained foreign bodies could cause serious neurosis as well as cardiac arrythmias; he believed that these were strong reasons for surgical treatment. But a patient who has harboured a foreign body in the heart for years without apparent harm will often not want it removed, and the surgeon may well hesitate to interfere, even though he knows that complications are likely in the end. Some of the late complications can still be resolved by the removal of a foreign body but many pass beyond surgical care.

CARDIAC TUMOURS

Primary Cardiac Tumours are rare, and account for about 0·05 per cent of autopsies. They can be important to the surgeon because if an accurate diagnosis could be made some might be removed: and because the condition may be encountered unexpectedly at cardiotomy. Three-quarters of cardiac tumours turn out to be benign, and half of these are myxomata. Myoma, fibroma and lipoma can occur in the septum, and some of these tumours can be shelled out of the heart without opening the chambers.

Myxomata usually grow from one of the atria, more often the left, in the vicinity of the septum near the foramen ovale. They are pedunculated jelly-like masses which can attain the size of an orange, attached to heart wall by a stalk which may enable the tumour to move about in the atrium. They are regarded as true tumours and not as organized thrombi; although the surface of the tumour may have a clot upon it.

Diagnosis. The site of the tumour determines the clinical picture produced in each case. A myxoma of the left auricle simulates mitral stenosis and leads to pulmonary hypertension, cardiac failure, and systemic emboli; the latter may be of clot or tumour fragments. Murmurs are variable or absent, and this point may suggest that the case is not one of mitral stenosis. A myxoma of the right auricle imitates vena caval obstruction without cardiac murmurs or arrhythmias. Relentless progress of cardiac failure, despite adequate obvious cause and despite rest and digitalization, is one of the diagnostic criteria. An abnormal outline on radiography has also been described. Innocent tumours do not interfere with the conduction mechanism, and electrocardiograms are normal. A myxoma situated in the right atrium may be outlined by dye injected into a peripheral vein, or by angiocardiography using a catheter. In the left atrium the diagnosis has most often been made at operation for supposed mitral stenosis. In such cases the mitral valve is normal to palpate and there should be no difficulty in distinguishing the features of a myxoma.

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d'Abreu has successfully removed a large simple hydatid cyst from the wall of the left ventricle. He removed another from the same patient, in the spleen, and the woman is well.

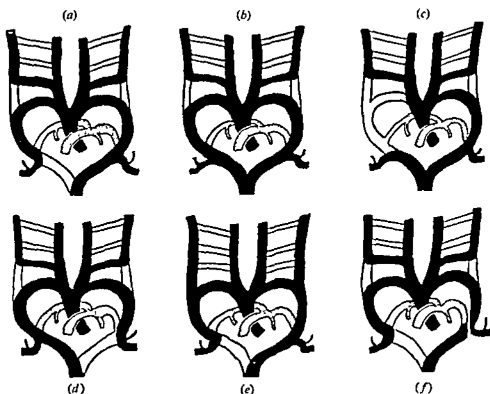
The treatment of hydatid disease in the heart is to take out the cyst intact, using the technique described by Barrett for shelling out pulmonary cysts.

CONGENITAL ABNORMALITIES OF THE AORTA AND THE PULMONARY ARTERIES

General Observations

The great vessels are derived, by a complex series of changes, from the primitive branchial arches. These evolutions occur according to a regular pattern and the final anatomy varies little in normal people.

In a minority anomalies occur and are due to the persistence of channels which should disappear, or to the disappearance of vessels which should remain. The variations which can occur upon this plan are legion: many cause no symptoms, but some are harmful and can be corrected. There are three ways in which an abnormality may cause trouble; it may constrict the œsophagus or the trachea, it may result in a shunt of blood from the systemic to the pulmonary system, or it may result in obstruction of an important blood channel.



(From Hamilton, Boyd and Mossman, "Human Embryology." W. Heffer and Sons.)

FIG. 273 Schemes to show abnormalities in the development of the Branchial Arch Arterial Pattern. Persisting systemic arteries are represented in solid black; the pulmonary trunk, its branches and the ductus arteriosus are stippled (a) Normal pattern (b) Double aortic arch (c) Abnormal origin of right subclavian artery. (d) Right aortic arch (e) Absence of both common carotid arteries (f) Extreme degree of coarctation of the aorta, the descending aorta being supplied by the ductus arteriosus alone

These deformities have been presented from time to time in the literature. At first they were described by physicians, such as Dr. Peacock, and by anatomists and pathologists; but recently surgeons such as Gross, Blalock, Murray, and Crafoord, have shown that many are curable. To understand the various combinations of abnormalities, and particularly to sort out unexpected findings at operation, the surgeon should have some knowledge of the prototype from which the adult anatomy of the great vessels has grown.



FIG 274 Complete transposition of the viscera The heart and the great vessels were normal

Surgical Embryology. The first evidence of a cardiovascular system is the appearance of two tubes which appear laterally within the mesodermol layer of the embryonic plate. Subsequently these tubes swing together and fuse to form a single heart lying ventral to the foregut and caudal to the buccopharyngeal membrane. At this stage the septum transversum forms; it lies caudo-ventral to the heart and separates the pericardium from the cœlom.

The cephalic end of the primitive heart extends forwards to the head beyond the pericardium as a tube, and terminates on each side in plexiform vessels growing in the

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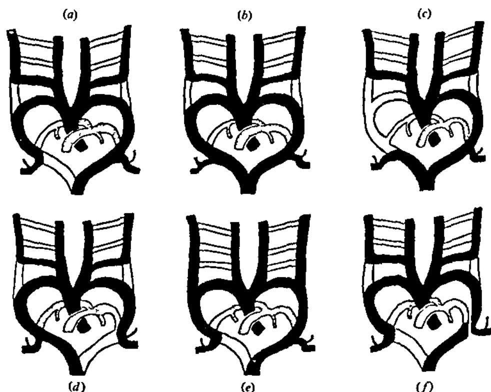
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The left innominate comes off first, then the right common carotid and finally the left subclavian. The aorta passes either behind, or in front of, the œsophagus and courses down the mediastinum slightly to the right of its normal position. The pulmonary vessels and the cavæ are normal, but many variations in the origins of the main arterial branches have been described. Most of these are of academic interest, because they cause

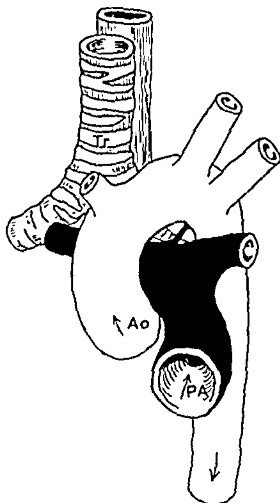


FIG. 275 Normal aortic arch.

NOTE Ligamentum arteriosum

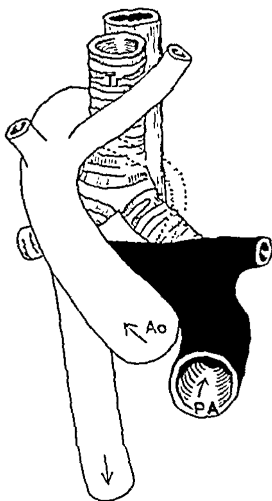


FIG 276 Right aortic arch Right descending aorta.

NOTE The ligamentum arteriosum may constrict the œsophagus and trachea if it follows the course shown in dotted lines (anomaly of Neuhauser).

no symptoms, and because surgical corrections are not necessary or possible. But some, by pressing upon the œsophagus or the trachea produce obstruction in infants. Others occur as associated abnormalities in various types of congenital cyanotic heart disease, and particularly in Fallot's tetralogy, and they add to the complexity of the various shunt operations devised to alleviate the pulmonary ischæmia. There are two main types.

A. The Anterior Type

In the anterior type the aorta crosses the right main bronchus and arches back over the front of the trachea. This is very rare but may be encountered in cases of transposition of the great vessels.

mesoderm of the mandibular arch. These vessels pass dorsally around the pharynx as the first branchial arteries and communicate with the two dorsal aortæ. As the heart develops in complexity it moves away from the neck and five more pairs of branchial arches form and connect the ventral to the dorsal arterial trunks. The caudal parts of the dorsal aortæ then fuse to produce a single trunk and this extends forwards but does not reach the pharynx. The 5th pair of arches are never fully developed in man, and the pulmonary arteries grow from parts of the 6th pair.

The dorsal aortæ between the third and fourth branchial arteries disappear as the heart "descends": and the segment of the right dorsal aorta, between the origin of the subclavian vessel and its junction with the left dorsal aorta, also goes. In mammals blood can only reach the dorsal aorta by traversing the remaining channels in the left fourth and sixth arches. The terminal segment of the left sixth arch persists, until respiration begins, as the ductus arteriosus: the terminal segment of the right side disappears, and the two proximal segments form the pulmonary arteries which enter the lung buds All these changes are complete in the human fœtus by the sixth week.

In fish and amphibia the branchial arteries carry the blood through the gills: in higher animals the branchial system is superseded, but the anatomical arrangements in early embryonic life are essentially the same in all vertebrates.

In a normal person those parts of the branchial arteries and of the aorta which disappear leave no anatomical trace. But it is not uncommon to find a fibrous cord between the pulmonary artery and the aortic arch in the position of the ductus arteriosus.

In unravelling the anatomy of these cases the surgeon is helped in two ways. The radiologist can often give an accurate picture by using angiocardiography and showing up the gullet with barium. Secondly, at operation, the exact anatomy of the vagus and the recurrent laryngeal nerves is decisive, because these nerves do not change their relationship to the fourth and sixth branchial arches as the adult anatomy of the aorta and the subclavian arteries evolves.

Dextrocardia

Dextrocardia is the condition in which the heart is rotated, so that the apex is directed towards the right side, and all other viscera are normal in position. The right ventricle lies in front of the left: the right atrium is to the left of the sternum and the left atrium is at the back.

Isolated dextrocardia without situs inversus is almost invariably associated with other congenital cardiac anomalies which may demand surgical treatment. Such cases present a formidable diagnostic problem but are fortunately very rare.

Dextrocardia with Situs Inversus

The heart and all other viscera lie on the opposite side of the body to the normal. It is a side for side reversal and does not cause abnormal symptoms referable to the cardiovascular system or to the viscera. It is rarely associated with cardiac deformities.

Both the above types of dextrocardia must be distinguished clinically from conditions which displace the heart from its normal position in the left chest.

ANOMALIES OF THE AORTIC ARCH

Right Aortic Arch. The aorta arises from the left ventricle in the normal way but arches over the right main bronchus. The anatomy of the three main vessels is as follows.

(2) The left subclavian artery may follow the normal pattern and arise from a persistent left aortic root which itself takes origin from the left dorsal aorta. In this case the aorta passes behind the œsophagus from right to left and the left subclavian artery lies in front (Fig. 278).

(3) Double aortic arch is a rare anomaly in which both aortic arches persist and then encircle the gullet and the trachea. It has often been missed because the surgeon is apt

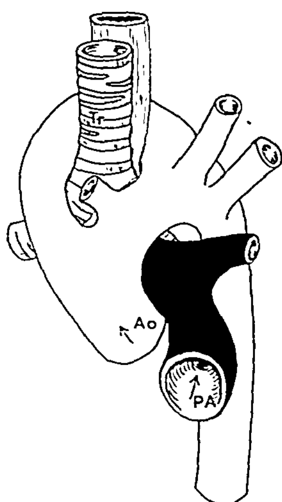


Fig 279 Double aortic arch

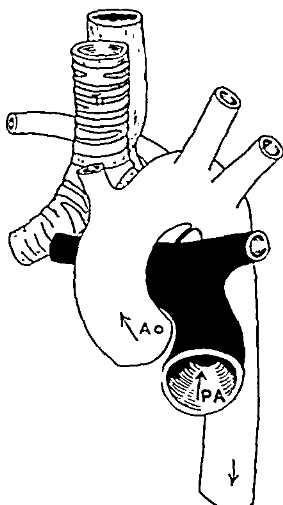


Fig 280 Normal aortic arch. Anomalous right subclavian artery.

Note Ligamentum arteriosum œsophageal compression.

to think of it in terms of two long vessels passing round the root of the heart like a necklace and joining together near the diaphragm. It looks nothing like this in life, because the double channel is confined to the region of the aortic arch. It may occur as two patent vessels of equal size, or a part of one may be obliterated as a fibrous ligament.

None of these varieties place any strain upon the cardiovascular system, and if they cause symptoms it is because the vascular collar, which encircles the œsophagus and the trachea, obstructs the gullet or the airway.

The Clinical Picture

The clinical picture produced by all types of vascular collar is the same: it is of a

B. The Posterior Type

The aorta passes to the right and then doubles back behind the œsophagus, and of this there are several variants according to the origins of the main branches. If the left subclavian artery, for instance, comes off to the right of the œsophagus and trachea these structures may be compressed as the artery passes to the left arm; relief from pressure

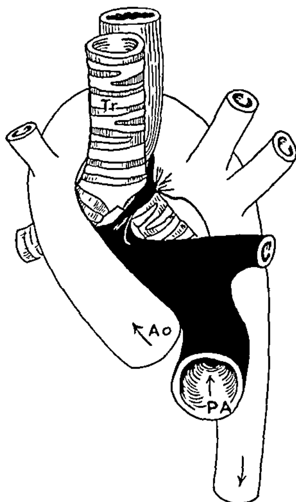


Fig 277 Right aortic arch. Left descending aorta. Ligamentum arteriosum constricts

Note Ligamentum arteriosum causing constriction

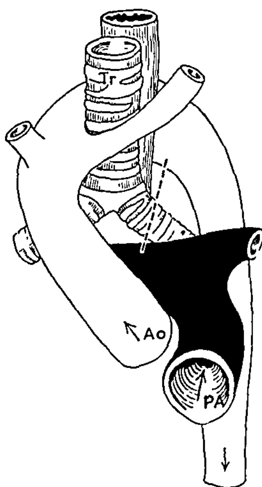


Fig. 278. Right aortic arch Left descending aorta.

Note Left subclavian artery causing compression The ligamentum arteriosum lies in the dotted line

symptoms would be afforded by dividing or transplanting the vessel. In some it is the ligamentum arteriosum which completes the collar encircling the œsophagus and trachea: division of this ligament relieves symptoms by breaking the ring.

Right Aortic Arch with Left Sided Descending Aorta. In some cases the aorta arches back on the right, as described above, but is then abruptly diverted to the left and descends in the normal place. When this happens *a part of the aorta lies behind the œsophagus* and causes an anterior displacement of the gullet and the trachea. This deformity often causes no symptoms, but in some cases it does and surgical treatment is possible. There are three varieties, which differ according to the reason why the aorta is drawn to the left.

(1) The ductus arteriosus or its vestigial ligament can draw the aorta to the left.

Surgical Pathology. Autopsy reports show that coarctation occurs about once in every 1,500 routine post mortems: this figure refers to occlusions which are complete or virtually so, but in many more patients a lesser degree of stricture is present. Coarctation is more common in men than women and occurs in all parts of the aorta, but the usual type results in a diaphragmatic pinhole stricture situated immediately distal to the ductus arteriosus. Sometimes an inch, or more, of the aorta is narrowed by the coarctation.

There are two varieties which affect the arch and these are called the "*infantile*" and the "*adult*." In the former the stricture involves the aorta anywhere between the origin

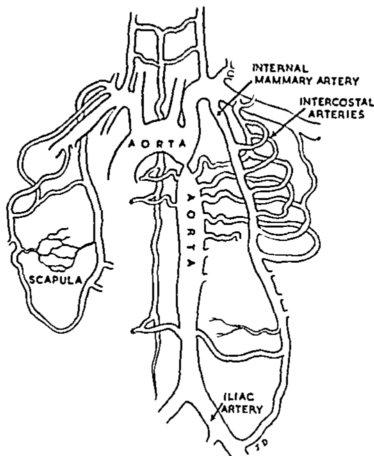


Fig. 281 Diagram to illustrate the chief collateral vessels which develop in a patient suffering from coarctation of the aorta.

Note. A diagram somewhat similar, but not identical with this appeared in *Proceedings of the Staff Meetings of the Mayo Clinic*, Jesse E Edwards *et al.*, p 333, July 21, 1948

of the coronary arteries and the ductus arteriosus: it is generally associated with multiple intracardiac anomalies and is incompatible with survival unless the ductus remains patent and carries blood to the systemic vessels. The "adult" variety is that which occurs

strictures are congenital abnormalities.

The external appearance of the aorta is often deceptively normal; but in most cases of constriction, there is an indentation of the aorta which is least deep at the site of the

sickly infant having attacks of croup and pulmonary infections. The attacks may date from birth so that congenital atresia of the œsophagus must be considered as an alternative diagnosis. The infant generally has wheezy respirations and an unproductive cough. During feeding stridor and cyanosis can occur. Dysphagia may be a prominent symptom but these babies can with patience, be fed, though they cough and splutter over every mouthful. Occasionally a case of this type occurs in an adult.

The Radiological Appearances

The radiological appearances which are diagnostic are these. There is a constant narrowing of the trachea opposite the third dorsal vertebra; there is no tracheal or œsophageal atresia. The œsophagus is compressed on all sides, and there are generally patches of pneumonitis in the lungs. If the condition is first revealed in an adult it may be because the part of the arch adjacent to the vertebral column has become aneurysmal, and, in this case, the bodies of the vertebræ may be eroded.

Treatment. The child can be cured by dividing the vascular collar at some point. The actual point selected depends upon whether the anterior or the posterior component of the aorta is larger, and upon the courses of the main branches *vis à vis* the gullet. d'Abreu stressed that in a patient upon whom he operated successfully the great vessels appeared to be normal on a first inspection: it was only after a great deal of dissection in the mediastinum that the true anatomy was disclosed.

Anomalous Right Subclavian Artery. This anomaly may cause symptoms identical with those of the anomalies of the aortic arch described above. The first case was described by Bayford; his patient was a woman suffering from dysphagia and his paper appeared in 1794. He considered the deformity to be a trick of nature and he called it

Dysphagia Lusoria. The condition generally causes no symptoms and is present in 1 in 200 autopsies. The right subclavian artery arises from the aorta beyond the origin of the left subclavian; that is, its origin is formed by the distal part of the right aortic arch which has persisted, instead of from the proximal reach. Holzappfel, who investigated 143 cases, described the anatomy as follows: the artery passed behind the œsophagus in 107, between the trachea and the œsophagus in 30, and in front of the trachea in 6: the beginning of the artery was often aneurysmal.

Treatment is only indicated to relieve dysphagia and this can be done by ligating and dividing the vessel. Neither gangrene, nor circulatory disturbances of the right arm need be anticipated.

COARCTATION OF THE AORTA

In coarctation the thoracic aorta is partially or completely occluded by a stricture. The fact that this stricture is often associated with bicuspid aortic valves makes it almost certain that coarctation is a congenital deformity of fœtal life. The "adult" type probably arises as a result of an extension of the tissue of the ductus arteriosus into the aorta, so that when this tissue contracts as the ductus obliterates, the aorta is also constricted. In such a case the coarctation might not be present at birth.

The deformity is frequently complicated by others such as bicuspid aortic valve, patent ductus arteriosus, anomalies of the origins of the branches of the aorta and of the cerebral vessels, cerebral aneurysms, and changes in the ribs and sternum. Subacute bacterial endarteritis of the coarctation site may complicate the picture.

aneurysmal, both at their junctions with the parent trunk, and anywhere along the intercostal groove, where they can cause "notching of the ribs." Sometimes the fine jet of blood which spurts through the pinhole impinges on a spot in the wall of the aorta beyond the obstruction, and by constant friction causes a weakness which acts as the apex of an aneurysm.

A generous collateral circulation develops between the vessels at the base of the neck,

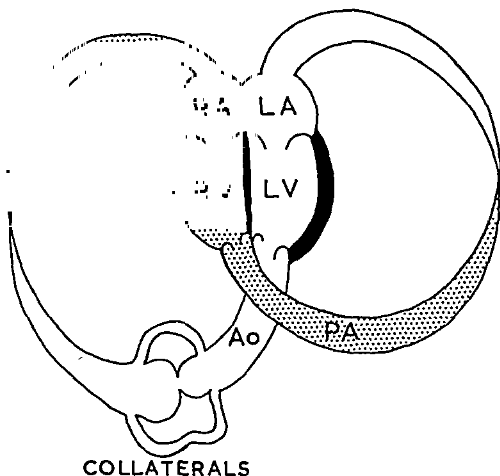


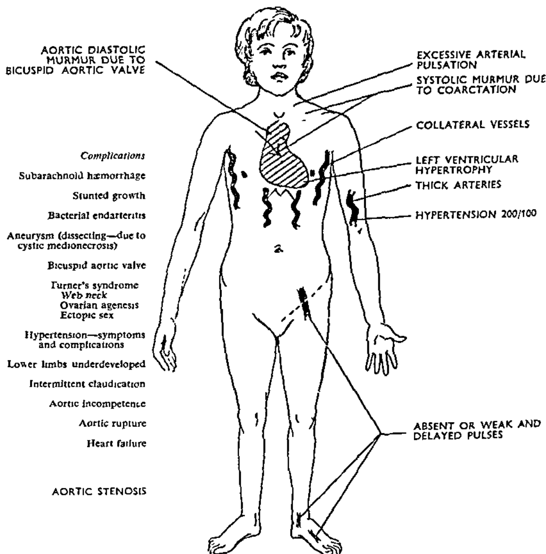
FIG. 283 Coarctation of aorta circulation.

NOTE Left ventricular hypertrophy. Abnormal aortic valves Post-stenotic dilatation of aorta. Collateral circulation

those which course around the pectoral girdle, the intercostals, the internal mammary and the epigastric vessels. Through these channels the blood passes to the lower limbs and the abdomen, and the majority of the typical signs are due to their presence.

The Diagnosis. Coarctation of the aorta frequently remains obscure and yet in most cases the diagnosis is not difficult if the surgeon is familiar with the symptoms and signs. The patients may complain of frontal headaches, palpitations, bleedings from the nose, and a feeling of congestion in the head. These symptoms are of hypertension. In the lower part of the body there are symptoms of claudication affecting the muscles of the legs or the buttocks, the femoral pulses are feeble or absent, and much delayed, t! peripheral circulation is not as good as that in the arms and the patient may notice that the legs get cold, or that chilblains are troublesome in winter. By contrast there are some

ligamentum arteriosum. The lower wall of the vessel is not indented, and into it the ductus or its ligament is inserted. The occlusion may be linear or it may involve a stretch of the aorta: the former is the more common. When the aorta has been opened the usual finding is of a structure like a diaphragm obstructing the lumen: in the lower and inner part of this diaphragm is the remaining channel which is seldom larger than 1 mm. in



PROGNOSIS Untreated average 33 years (Abbott's series of 68 cases) Treated—hypertension relieved, eventual result uncertain complete success 98 per cent (Gross) Operative mortality—5–10 per cent

FIG 282. Coarctation of aorta

diameter. It is not possible to assess the degree of stenosis by looking at the outside of the vessel. Histologically the lesion consists of fibrosis with replacement of the media by scar and there is thickening of the intima in the vicinity. Proximal to the stricture the wall of the aorta is hypoplastic, the intima often shows patches of atheroma and the media may be partially calcified. Immediately distal to the obstruction the aorta is larger than normal and into it open pairs of dilated, tortuous, anastomotic, intercostal vessels carrying the blood from the chest wall back into the aorta These intercostals are often

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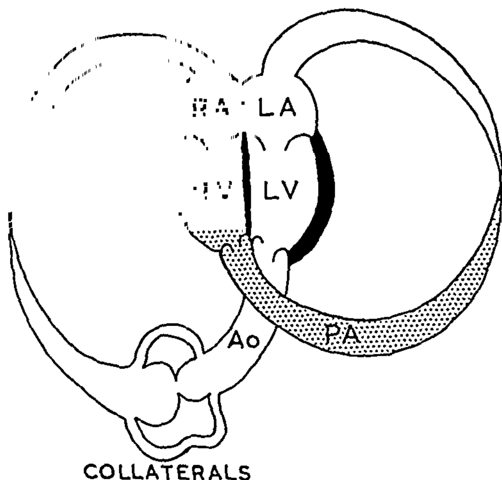


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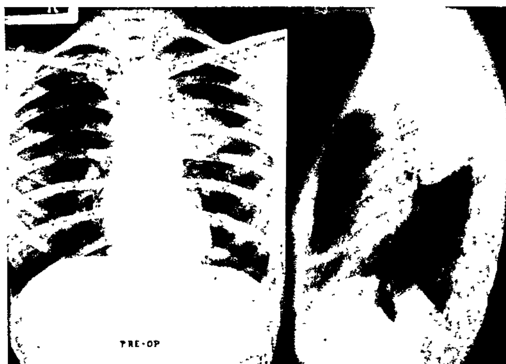


FIG 284. Coarctation of the aorta with an aneurysm of the descending aorta situated immediately distal to the coarctation.



FIG. 285. Tomogram showing the aneurysm in which there are areas of calcification.

patients who have gross coarctation and who are symptomless, whilst others are principally worried by gastro-intestinal upsets. The harmful effects do not always seem to be progressive.

One of the most important aspects of coarctation concerns the blood pressures and the pulses, and about these some confusion exists. The position is this. The pulses in the lower limbs are delayed and difficult to palpate. The blood pressure in all branches of



Fig 286 Aortogram showing that the shadow seen in the plain radiogram is, in fact, an aneurysm

the aorta above the coarctation is raised. Electromanometric pressures taken from the femoral artery show that the pulse pressure is lower in the leg than in the arm, but the mean pressure is not significantly changed. This is because the pulse wave is damped by the passage of the blood through the collaterals. The pulse in the legs may be normal in spite of coarctation if efficient collaterals are present.

When the collateral vessels are well developed, pulsations can be felt in many arteries which are not normally palpable. In particular those which course along the vertebral borders of the scapulæ can often be seen and palpated. The blood passing through these large collaterals may give rise to systolic murmurs along the course of the vessels. But murmurs are variable in coarctation. If the arterial block is *complete there will* be no



FIG 287 The aneurysm was excised and the gap in the aorta was bridged with a piece of human aorta which had been stored by freeze-drying for several months.



FIG. 288. Specimen cut open to show the various pockets, zones of calcification and the pin-point openings of the aorta into the aneurysimal sac.

murmur directly due to the obstruction, but if a small jet remains in the aorta there may be a systolic thrill or murmur, best heard over the back of the left upper part of the chest, or to the left of the spine. A mid-diastolic murmur is not uncommon, and is due to aortic incompetence resulting from a bicuspid aortic valve.

On radiography the heart is often of normal size, but the left ventricle is usually hypertrophied: the aortic knuckle is small and above it the dilated left subclavian artery

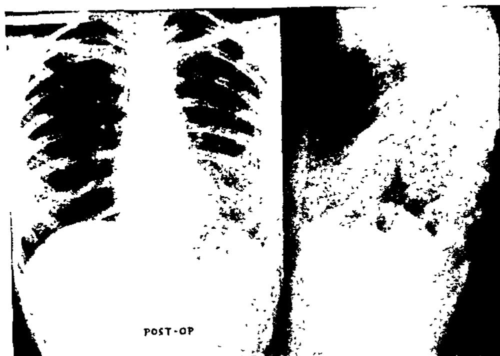


FIG. 289 Two years later the patient is well, the pulses in the legs are normal and there is no abnormality in the chest X-rays

forms a crescentric bulge upon the left side of the upper mediastinal shadow. The ribs may be notched but this sign is not always present; it is not pathognomonic of coarctation for other vascular lesions can do the same. Tomograms of the aortic arch region may be helpful in depicting areas of calcification. In one quarter of all cases there is no direct radiological evidence of coarctation.

The electrocardiogram usually gives evidence of left ventricular hypertrophy.

The diagnosis can be confirmed by aortography done in such a way that the tip of the catheter, introduced retrograde either up the brachial or radial artery, lies adjacent to the stricture. In some cases good definition of the anatomy can be had by introducing the dye into a vein and waiting for it to reach the systemic vessels. The anatomy of the coarctation, which must be known before operation, can only be decided by angiocardiography; but this investigation gives the surgeon no idea of the state of the aortic wall itself

The Prognosis. In the "adult" type the prognosis is variable, for it has been shown that the signs and symptoms are not directly proportional to the size of the lumen of the coarctation. Nor is the mechanism of "L-"

aortic obstruction: it may be that the r

a lack of pulsatile blood supply to the kidneys. Suffice to say that two patients having apparently similar lesions may not behave in the same way. On the other hand there is no doubt that the lesion is generally dangerous to life if it is associated with demonstrable signs and symptoms. The following complications can occur: cerebral hæmorrhage, cardiac failure, rupture of the aorta, a localized or a dissecting aneurysm, bacterial endarteritis, and fibrosis of the endocardium of the left ventricle. Blackford, who studied 196 autopsy reports, stated that 40 per cent of the patients died between the ages of 16 and 30 years of age. In Abbott's series, comprising 68 patients, the mean age of death was 33 years (page 554).

Treatment. Three different surgical interventions have been advocated. Cervical sympathectomy has been shown to be of no avail. Secondly it was suggested that the obstruction might be "by-passed" by turning down the dilated left subclavian artery and anastomosing it to the descending aorta; this has been tried and discarded because it cuts off many of the collaterals and does not get any more blood to the trunk than before operation. The successful results have come from resecting the coarctation and doing an end to end anastomosis of the aorta or bridging the gap with an arterial graft. The feasibility of this procedure was proved experimentally by Gross and Hufnagel in Boston; the first successful operation upon a patient was performed by Crafoord of Stockholm, and the procedure has now become generally accepted and widely practised. At first it was assumed that to clamp the thoracic aorta sufficiently long to do an anastomosis would be fatal to the spinal cord and the kidneys. But in coarctation the aorta can be clamped indefinitely because the collaterals carry the necessary blood. In human subjects, the normal aorta can only be clamped in the thorax for a few minutes. Another fear was that the anastomosis would not be sufficiently strong or secure to prevent fatal bleeding. It is a fact that a sound anastomosis can be achieved.

The Risks of Conservative and Surgical Treatments. Gross (1947) and his colleagues noted that, in 104 patients who were found at autopsy to have coarctation, the length of life had been as follows. 26 per cent lived a normal life and died from other causes; 74 per cent died as a result of the coarctation. The causes of death in this 74 per cent were—bacterial endocarditis 22 per cent; aortic rupture 23 per cent, and 29 per cent a result of hypertension The average age at which these patients died was 30 years.

The mortality, directly due to operations, varies in the hands of different surgeons but the following figures have been quoted: Clagett (1951) 70 cases: 7·1 per cent mortality; Gross (1953) 270 cases with 15 deaths in the first 100, and 2 deaths in the last 100. The final result of operation, according to Gross, is "complete success" in 98 per cent of cases.

The surgeon will thus have to equate the dangers of conservative and surgical measures, according to his own skill. This operation is not easy to perform until one has had a considerable experience of arterial surgery, but the anastomosis can be practised in animals before operating on a patient.

Selection of Patients for Operation. A patient who has a coarctation and who has symptoms which are progressive should undergo operation; but in this and all other types of cardiac surgery practice improves results, so that the risks will be high in the hands of those who are inexperienced. Resection of a coarctation is probably the most dramatic and the most potentially frightening operation in surgery today; it may take a long time to do and at each stage it presents difficulties and dangers.

Most surgeons prefer to operate upon children between the ages of 10 and 15. In them the vessels suture well and a gap of a centimetre or two can be bridged without undue tension. Over the age of 30 the proximal aorta is generally pathological and holds stitches badly; but successes have been recorded in patients over 50 and some of these have had a sound aorta at that time. Many patients over the age of 25 have renal changes producing permanent hypertension and they will not be cured by surgical treatment.

Technique. The best approach to the descending aorta is by a posterolateral thoracotomy. The first difficulty the surgeon encounters is in dealing with the plethora of anastomotic vessels in the muscles of the chest wall. Every one of these must be meticulously ligated and hypotensive anæsthesia is invaluable. The pleura is opened by a generous incision through the bed of the fourth rib. It is better not to make an intercostal incision because of the danger of wounding the intercostal vessel which may be aneurysmal. The lung is retracted and the pleura over the aorta is reflected. The next step is to mobilize the area of the coarctation and to free a sufficient stretch of the aorta on either side to allow approximation after excision of the stricture. Here the danger is that in freeing the aorta one of the large, delicate, intercostals will be torn and bleeding, which can be difficult to control, will occur from both ends. If the torn vessel is entering the aorta on the far side the surgeon may have created an almost insoluble problem. To avoid this, patience and absolute gentleness in dissection are required. Having mobilized the coarctation a decision must be made as to whether any of the intercostal collaterals must be divided; on the whole the more the intercostals can be saved the better. Two special arterial clamps are now applied and the stricture is excised. After excision some tension may be necessary to get the ends together but in children, and after proper mobilization, this is not a difficulty in most cases. A greater problem is that the lumen of the proximal part may not be as great as that of the distal, and some adjustment may be necessary on this account. Before attempting to sew the ends together the adventitia must be completely removed: if this is not done the stitches get tangled and do not run easily through the tissues. The result is that unnecessary holes are made and these cause difficulty later in the operation. The anastomosis should be done with fine arterial silk sutures, using a continuous everting stitch. Only one layer is desirable, and if, when the clamps have been removed, the suture line leaks a little, it is better to apply prolonged pressure rather than to cobble it up with numerous interrupted stitches. If the suture line leaks badly it is necessary to excise the ends and start again. To achieve a good result the resulting lumen must be at least 80 per cent of the normal for that patient. When the suture line has been completed the clamps should be slowly loosened over a period of minutes in order not to deplete the flow of blood through the coronary arteries too quickly. The pleural cavity should be drained for 24 hours in order to reveal any leak of blood in the reactionary period.

If, during the course of the operation, serious bleeding occurs the surgeon should deal with the situation and postpone any further intervention for a period of 10–14 days. Some of the most successful operations have been done in two stages.

Arterial Grafting. In some cases it happens that the stricture is so long that if it were to be excised the ends could not possibly be brought into apposition for anastomosis. To meet this possibility, which can generally be anticipated by proper aortography, Brock thinks that a coarctation should never be undertaken by a surgeon who is not prepared to bridge the gap with an arterial graft. This involves having such a graft ready

and available on every occasion. In spite of this advice hundreds of operations have been done by surgeons who had no grafts at hand; but the ideal put forward by Brock is, without doubt, sound. Grafting these cases was introduced by Gross, and its worth has been amply proved.

In 1908 Carrel, and in 1912 Guthrie, proved that homologous vascular grafts could be used to restore arterial defects. In recent years interest in this subject has revived and a great deal of work on how to store homologous grafts has been done. Broadly speaking two proved methods of storing grafts are available.

In the one, advocated by Gross, the object is to secure a graft in which the cells are still alive at the time it is implanted. To achieve this, segments of arteries, removed from the bodies of subjects recently dead, are kept sterile and are stored in a buffered nutrient saline solution at — 4°C. Such grafts live for 4–6 weeks and can be implanted at any moment within this period. Gross has successfully applied this technique to more than 50 patients, suffering from coarctation, in whom he has excised the stricture and bridged the gap with a human aortic graft.

The second way is to freeze the graft. This has been carefully studied by many workers but particularly by Hufnagel and Eastcott. The object is to freeze the individual cells of the graft in such a way that they are not disrupted: that is, a piece of tissue treated in this way appears to be normal when examined histologically but is, in fact, dead. Freezing does not kill all organisms and has no effect upon viruses so it is not a method of sterilization in itself. The graft must still be taken from the donor under full aseptic precautions.

Grafts can be frozen either at — 70°C. using solid CO_2, or at — 195°C. using liquid nitrogen and kept in a "deep freeze" indefinitely. Tissue injury from ice crystals within the cells varies inversely with the speeds of freezing and thawing, and chemical changes are prevented by low temperatures. Crystallization can be prevented altogether and the cells in the graft remain intact for any length of time.

The most convenient way in which to store grafts of this type is to use a combination of freezing and drying. The graft is secured in the ordinary way and frozen: it is then put into a glass tube from which the air is withdrawn. In this form it can, apparently, be kept for any length of time in any climatic conditions.

The ultimate fate of all types of homologous aortic grafts is degeneration and fibrous tissue replacement by the host. It has been proved that these grafts supply a scaffold for a new host intima and adventitia, and that they function as satisfactory pipes for the transmission of blood during the healing period. Experience has shown experimentally, and in human operations, that the principal failures occur within a few days of inserting the grafts and are due to thrombosis or disruption. It has been proved that live grafts and dead grafts usually survive longer than this danger period; most surgeons who use "live grafts" do so because they believe the endothelium actually takes part in the healing process before it disintegrates.

Many different techniques of collecting, sterilizing, and storing grafts are under trial. It has, for instance, been found possible to sterilize material taken without aseptic precautions from the autopsy room, by exposing it to cathode rays. Some special organization is necessary to store grafts so that they may be available at the required moment.

During the last 2 years many synthetic materials, such as nylon, orlon, etc., have been

used successfully for bridging defects in large arteries. These materials can be cut and tailored to the required shape and sterilized by boiling. The only disadvantage that has been found is that, if used in a child, the lumen of the graft does not grow as the child grows. Plastic grafts are particularly valuable as temporary by-passes.

Autogenous and synthetic grafts heal well, and seldom give rise to aneurysmal dilation or rupture. The great merit of grafting is that it enables the surgeon to bridge a gap of any length without tension and, if a large graft is used, a normal lumen can always be achieved.

Results. Resection of a coarctation is difficult, exacting and dangerous: nevertheless the necessary techniques can be mastered in animal experiments by competent surgeons. The operative mortality should not exceed 15 per cent, and, with practice, it is less. The immediate causes of death are primary hæmorrhage due to tearing of a hypoplastic aorta, intractable bleeding from an intercostal vessel, or cardiac arrest after the clamps have been removed. If the operation goes well convalescence should be uneventful, but bleeding from an injured intercostal has occurred up to 5 weeks after the operation. Cerebral accidents and embolism also occur. In general the results of successful surgery are excellent; the symptoms are relieved and the patients develop normally. In some the blood pressure takes a long time to return to normal. It may be that the hypertension persists because of permanent changes in the renal vessels; or because all the vessels arising below the original coarctation are diffusely hypoplastic. Clinical improvement can occur without complete normality of the blood supply to the extremities.

Aortograms, done some years after operation have proved that the diameter of the aorta at the suture line grows as the child gets older.

PATENT DUCTUS ARTERIOSUS

The ductus arteriosus is the remains of the outer part of the sixth branchial arch on the left side. During fœtal life it is the channel by which the blood passes from the pulmonary artery into the systemic circulation, by-passing the lungs.

Surgical Anatomy. The ductus connects the left pulmonary artery with the inner side of the aortic arch. It passes from a point opposite the origin of the left subclavian artery to the top of the left pulmonary artery near to its origin. In direction it travels upwards, outwards, and backwards from the pulmonary artery. On its medial side, in the gap between the aorta and the pulmonary artery, is a minute gland similar in structure to the carotid body. Behind the ductus is the left main bronchus. The left recurrent laryngeal nerve, which comes off the vagus as it reaches the top of the aortic arch, diverges gradually from the vagus, winds round the aorta, and touches the outer side of the ductus. This



Fig. 290. Th...

nerve is the principal surgical landmark in identifying the ductus. In front of the ductus there is an upward reaching pouch of pericardium and in many patients there are enlarged lymph glands in front of the pericardium. The ductus cannot be seen until the mediastinal pleura, and the pericardial pouch in front of it, have been reflected.

The ductus arteriosus is thin and can be obliterated by the gentlest digital compression. One might have expected a patent ductus to behave as do other arterio-venous fistulæ;

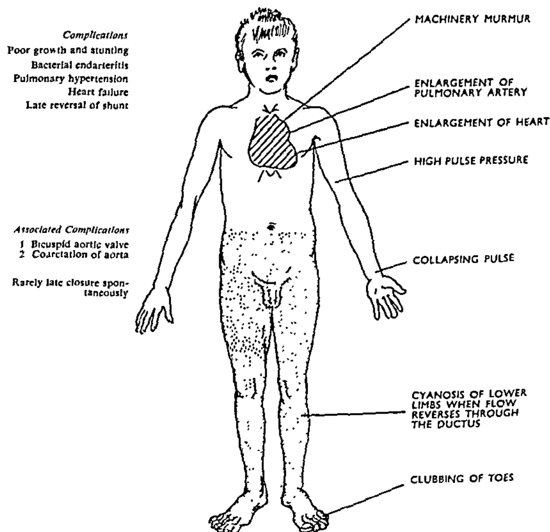
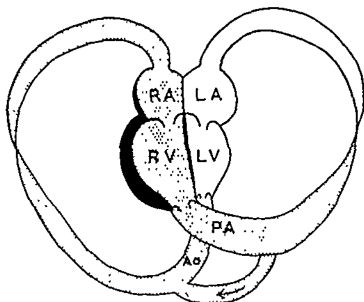


FIG 291. Patent ductus arteriosus with reversed shunt.

that is, the pulmonary artery should be "arterialized" and distended. But in this defect the blood does not always flow from the aorta into the pulmonary artery and it is probable that in many uncomplicated cases the shunt may be minimal. There are physical reasons which support this idea; the direction in which the opening of the ductus faces the aortic stream varies, and the opening in any case lies upon the concave side of the arch where the flow is less and eddies occur In a few who suffer from pathological hypertension in the lung vessels, the blood flows permanently from the pulmonary artery to the aorta: in these patients there is cyanosis of the lower parts of the body, polycythæmia, and clubbing

of the toes. In children these last signs can also occur as a result of a ductus which enters the aorta below a coarctation. The ductus can become aneurysmal and cardiac enlargement often occurs: this is the result of the increased work which both ventricles have to perform.

Normal Closure. The ductus probably closes as a result of oxygenation of the blood, which occurs as soon as the infant breathes. The exact method of closure is disputed but the muscle in the wall of the vessel contracts and becomes œdematous. The fibrosis,



LATE STAGE—right to left shunt with right ventricular hypertrophy and pulmonary artery hypertension

FIG. 292 Patent ductus circulation Late changes.

which subsequently forms the ligament, is probably the result of organization of this œdema fluid.

TIME OF CLOSURE

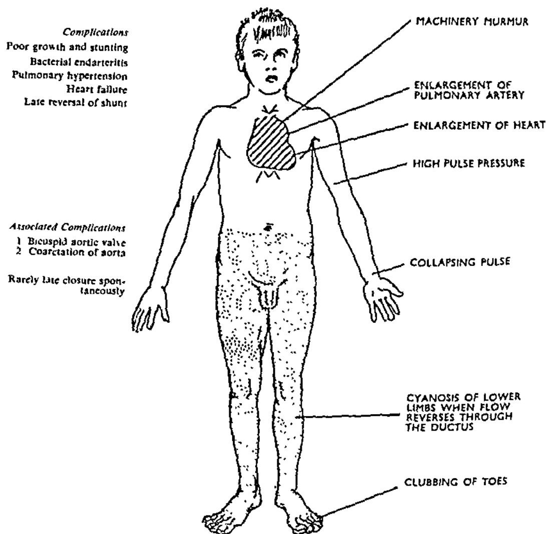
The ductus should close physiologically immediately after birth and anatomically at any time during the first 3 months of life. In most adults it remains as a fibrous cord called the ligamentum arteriosum; but in many the vestige of a lumen persists although blood does not circulate through it. If a patient at any age after infancy has the physical signs of a patent ductus it is a safe assumption that it will not close.

THE HARMFUL EFFECTS OF A PATENT DUCTUS

The presence of a shunt from the aorta into the pulmonary artery, where the pressure is lower, is a danger to the patient. The reasons for this danger are these. If the ductus is large so much blood may be diverted from the systemic circuit that growth is retarded, or the left side of the heart may be unable to maintain the necessary additional output and congestive failure occurs. The presence of a patent ductus is a cause of bacterial arteritis, especially of the variety due to streptococcus viridans, and the normal expectation of life is halved. But a person with a patent ductus may die of old age, never having been ill on that account.

nerve is the principal surgical landmark in identifying the ductus. In front of the ductus there is an upward reaching pouch of pericardium and in many patients there are enlarged lymph glands in front of the pericardium. The ductus cannot be seen until the mediastinal pleura, and the pericardial pouch in front of it, have been reflected.

The ductus arteriosus is thin and can be obliterated by the gentlest digital compression. One might have expected a patent ductus to behave as do other arterio-venous fistulæ;



These are the cases in which permanent pulmonary hypertension is present and operative closure of the ductus is dangerous

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earlier than this if complications or heart failure are already present. If the ductus has become infected operation must be deferred until a full course of antibiotics has been given and the infection has been controlled. Operations carried out in the presence of bacterial endarteritis carry a relatively high mortality and only cure about 60 per cent of the cases. A congenital defect of the aortic valve, which is occasionally associated with this lesion, is no bar to success; but the operation may not be wise if complicated by an inter-ventricular septal defect. Special care must be taken in advising patients who have a patent ductus and pulmonary hypertension: if the lung changes are permanent ligation of the ductus will do harm. At the moment the selection and treatment of this last group of patients is "sub judice."

One final point requires emphasis. The loudness of the murmur bears no relation to the size of the ductus or to the amount of blood passing through it.

Surgical Treatment. The simplest, and in the opinion of many surgeons, the best operation is ligation of the ductus in continuity, placing a ligature at each end. If the ductus is properly ligatured and the lumen completely obliterated the risk of recanalization is small. But Gross, whose experience in this matter exceeds that of other surgeons, prefers to divide the ductus and to close the aorta and the pulmonary artery with stitches. This operation takes longer than ligation in continuity, and carries a greater risk: nevertheless many surgeons of experience prefer and advise it.

Technique. The operation is done under general anæsthesia and the left pleural cavity is opened by a postero-lateral intercostal incision; or by a vertical axillary incision as advocated by Denis Browne. The lung is retracted and the mediastinum exposed: an incision 3 in. long is made through the mediastinal pleura from the left subclavian artery on to the descending aorta. The edges of the pleura so cut are elevated and by dissecting in the loose areolar tissue in front of the arch of the aorta, the vagus nerve is carried forwards in the flap out of harm's way. The left recurrent laryngeal nerve is identified; it must be preserved from injury or inclusion in ligatures: it dives backwards immediately lateral to the ductus between the aorta and the left pulmonary artery. The upper surface of the left pulmonary artery is cleaned and the adventitia dissected from the front of the aortic arch. The pericardial sac in front of the ductus is dissected free and turned down. Unless this sac is dissected off the ductus accurate definition of the latter is impossible It is at this stage of the operation that the surgeon is able to assess whether he is dealing with a straightforward case, in which no special difficulties need be anticipated, or whether some complicating factor such as an aneurysm of the ductus, or of the aorta, or a partial coarctation is present. It is not uncommon to find an indentation of the aorta, without functional coarctation, in patients who have a simple patent ductus arteriosus

If difficulty is anticipated in dealing with the ductus, the arch of the aorta, in the vicinity of the ductus, must be mobilized so that complete control may be available if bleeding should occur. Arterial clamps must be available. When the front and the lateral parts have been defined the next step is to dissect the back of the ductus off the left main bronchus This is the dangerous part of the operation and is done most safely by keeping, as far as possible, against the back of the aorta rather than against the ductus itself. A pair of curved, blunt-nosed, artery forceps is the safest instrument to use. If, at this stage, hæmorrhage occurs from behind the ductus the area must be packed for 15 minutes and sometimes the bleeding stops; if it does not, the arch of the aorta and the

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earlier than this if complications or heart failure are already present. If the ductus has become infected operation must be deferred until a full course of antibiotics has been given and the infection has been controlled. Operations carried out in the presence of bacterial endarteritis carry a relatively high mortality and only cure about 60 per cent of the cases. A congenital defect of the aortic valve, which is occasionally associated with this lesion, is no bar to success; but the operation may not be wise if complicated by an inter-ventricular septal defect. Special care must be taken in advising patients who have a patent ductus and pulmonary hypertension: if the lung changes are permanent ligation of the ductus will do harm. At the moment the selection and treatment of this last group of patients is "sub judice."

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left pulmonary artery must be mobilized so that both may be clamped on either side of the ductus until control has been regained. In most cases the back of the ductus can be safely mobilized from the front of the left main bronchus by blunt dissection, and the vessel ligated in continuity. The technique of ligation varies: some use one stout ligature, whilst others prefer two ligatures placed at the ends of the vessel and as close to the aorta and the pulmonary artery as possible. These are matters of individual preference. Having ligated the ductus the thrill, which is present in the main pulmonary artery at some little distance from the origin of the ductus, disappears, and the machinery murmur is replaced by a soft systolic bruit.

If the surgeon elects to divide the ductus and oversew the ends, a different technique is necessary. Gross uses two narrow arterial clamps: these are placed at each end of the ductus and the vessel is then divided. The clamps do not crush the tissues and the ends are closed by continuous fine arterial stitches. Only one layer of stitches is necessary. To do this Gross does not mobilize the arch of the aorta and the pulmonary artery; but a surgeon who is less familiar with the possible hazards associated with having the aorta and the pulmonary artery open at the same time, may think it wise to do so.

Some advise pleural drainage as a safety measure after operation, but many find this step unnecessary. There is no special postoperative treatment.

The Prognosis. The mortality of operations done before the age of puberty is about 2 per cent or less, and deaths have been due to technical faults and mistaken diagnoses.

Gross and Longino (1951) reported on a series of 412 operations upon the ductus; the mortality was 2·1 per cent; but in patients who had no complications prior to surgery it was 0·5 per cent. H. W. Scott (1950) reported 180 cases of whom 5 died, and in whom there was one recanalization in which the ductus had been treated by simple ligation. 161 of the patients were treated by division of the ductus and suture and in this group there were 2 deaths. The operations performed by these surgeons have cured at least 98 per cent of patients.

The only late complication of importance is recanalization of the ductus and return of the machinery murmur. If this is going to happen it generally does so within the first 6 months after operation. There is no agreement as to whether recanalization is due to faulty technique at operation or to some natural healing process. It has occurred after ligation and after division of the ductus, but is rare in the latter event. It occurs less frequently as the experience of the surgeon increases, and most commonly in cases where the ductus has been very large and thin, or the operation difficult. Operations done for recanalization can be difficult and dangerous.

Patent Ductus Arteriosus Complicated by Bacterial Arteritis

This complication which used to be fatal is now rare because of the efficiency of the antibiotics. Before chemotherapy or antibiotics were introduced, Tubbs, working at St. Bartholomew's, showed that ligation of the ductus generally cured the infection immediately, and that the patients could be saved by operation alone. Nowadays if a patient, who has not been treated surgically, develops arteritis the infection is first controlled by penicillin or other antibiotic and the ductus is then ligated. The mortality in this group is higher than in the uncomplicated cases, because the ductus is often inflamed and the surrounding tissues may be œdematous.

Patent Ductus Arteriosus and Pulmonary Hypertension

The ætiology of pulmonary hypertension combined with patent ductus arteriosus is at present unknown, and the pathological changes found in the pulmonary vessels are not constant. The pressure in the pulmonary artery is as high or higher than that in the aorta and the shunt may be reversed; thus the blood in the pulmonary artery flows into the systemic circulation and the lower half of the body becomes cyanosed. Ligation of the ductus will cure the patient provided the pulmonary hypertension is not due to permanent abnormalities of the arteries in the lung. If these vessels are sclerotic or incapable of modifying their internal capacity, ligation of the ductus produces acute right-sided heart failure, and for this reason, it is advisable to clamp the ductus temporarily before applying permanent ligatures and then, by observing events and measuring the pressures in the aorta and the pulmonary arteries, to assess whether the patient can tolerate ligation.

ANOMALOUS PULMONARY AND SYSTEMIC VENOUS DRAINAGE

Anomalous venous drainage may be of importance not only in patients suffering from atrial septal defects, but in other ways. The cases may be divided into three groups. Anomalies of the systemic veins alone, those of the pulmonary veins, and those in which both the systemic and the pulmonary veins are associated with pulmonary systemic communications.

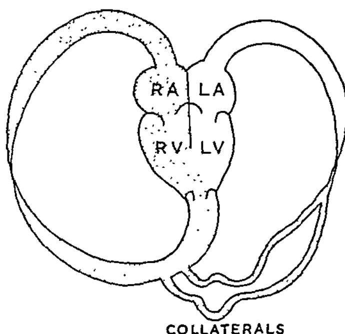
(1) *Systemic veins.* The commonest is persistence of the left superior vena cava. The left cava may drain into the coronary sinus and it may connect with the left innominate vein. Alternatively it may receive one or more pulmonary veins. These deformities are usually associated with other cardiac anomalies such as Fallot's tetralogy, aorto-pulmonary septal defects, coarctation of the aorta and pulmonary stenosis.

(2) *Pulmonary veins.* The number of these vessels may be unusual, or one, or more may drain into the vena cava or to some unusual part of the atrium. These anomalies are important to the surgeon in two types of case. Anomalous venous drainage complicates the operations done to close atrial septal defects. Secondly congenital stenosis of the pulmonary vein causes the chamber of the left atrium to be divided into two parts by a septum. Although there is a hole in this septum, it obstructs the return of blood from the lungs and produces symptoms like mitral stenosis. This anomaly can be easily cured by dividing the septum It is called Cor Triatrium.

(3) *Anomalies involving pulmonary-systemic venous communication.* In these cases there is an associated arteriovenous shunt which may or may not be complicated by a veno-arterial shunt elsewhere. They cause important changes in the mechanics of the circulation and influence the surgical treatment of some patients suffering from conditions such as atrial septal defects.

These deformities cannot be diagnosed with certainty before operation. They may be suspected if unusual dilatation of the right superior vena cava is noted in X-rays, if a left superior cava can be seen, or if there are curved shadows along the left or the right heart border. Confirmation can be had sometimes from cardiac catheterization which can not only show the catheter passing into an abnormal vein, but can demonstrate the presence of intra-cardiac shunts.

Type I. Pseudo truncus.

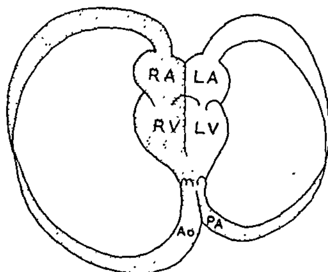


Lungs vascularized by collaterals

NOTE. Increasing the number of collaterals by performing pleurodesis may help

FIG. 293. Pseudotruncus arteriosus.

Type II. True truncus.



Lungs vascularized by pulmonary arteries arising from common arterial trunk.

FIG. 294. Aorta-pulmonary defect

PERSISTENT TRUNCUS ARTERIOSUS

In this condition the bulbus cordis has not divided into separate pulmonary and aortic vessels. Two main varieties exist, one of which is amenable to successful surgical treatment and the defects in the other may be somewhat alleviated.

Type 1.

MORBID ANATOMY (*Pseudotruncus arteriosus*)

The aorta and the pulmonary artery form a common stem which itself springs from both ventricles. In such cases the blood, destined to circulate through the lungs, either reaches them by way of two stout pulmonary branches arising from the arch of the aorta, or through a number of intercostal arteries derived from the descending aorta. In either case the whole of the circulating blood does not pass through the lungs, and these structures stand in relation to the systemic circulation in much the same position as do the kidneys. Such a condition is compatible with reasonable health and there is, in the museum at St. Thomas's Hospital, a specimen removed from a soldier who had been doing full military duties and who dropped dead on parade.

The diagnosis is difficult to make on clinical grounds alone. The patients are often discovered in childhood, either because they have signs of progressive limitation of exercise tolerance or because they are thought to be suffering from Fallot's tetralogy. The most important diagnostic points may be revealed by cardiac catheterization, angiocardiography and estimation of the oxygen saturation of the arterial blood.

Treatment. No curative treatment has been devised; but the operation of pleurodesis can help to get more of the systemic blood through the lungs (q.v.).

Type 2.

MORBID ANATOMY (*Aorto-pulmonary defect*)

The septum between the pulmonary artery and the aorta has partially developed; the two great vessels arise from the correct ventricles and are separate, but a foramen of varying size persists between the ascending aorta and some part of the pulmonary arterial system. The result of such a defect is that much blood from the aorta is shunted back and recirculated through the lungs unnecessarily.

Diagnosis. The importance of this condition to the surgeon is that the signs and symptoms imitate those of a patent ductus arteriosus. But, the machinery murmur is heard a little more to the right of the chest and other details may suggest the correct diagnosis. Whenever such a lesion is suspected full catheter and radiological studies are necessary before contemplating operation; for, whereas closure of a patent ductus is a relatively simple matter, closure of an aorto-pulmonary defect is difficult and dangerous.

The lesion must also be distinguished from an aneurysm of a sinus of Valsalva which has ruptured into the right ventricle; and from anomalous communications between the pulmonary and the coronary arteries.

Treatment. It is possible to close the defect in some cases. To do this it is either necessary to dissect out the connection between the two vessels, and obliterate it as one would a patent ductus arteriosus; or, using cooling or other technique, to open one of the great vessels and suture the aperture from within. The latter technique is generally agreed to be the better of the two.

CONGENITAL HEART DISEASE

Introduction

More than a hundred different congenital deformities of the heart have been described; this chapter concerns those which are compatible with life and produce disabilities which can be partially or completely corrected by surgical operations.

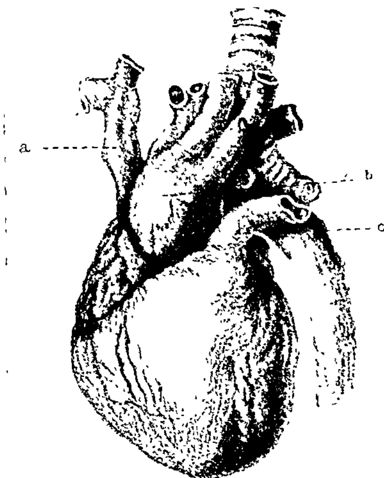


FIG 295 Lithograph taken from the book by Thomas Peacock (1858) on Congenital Malformations of the Heart. It shows the complexity of the deformities which may exist and hence the need for accurate pre-operative diagnosis In this specimen "a" is the superior cava, "b" the aorta and "c" the pulmonary artery The left ventricle was small and thin The aorta was large and arose from the sinus of the right ventricle which was hypertrophied and communicated with the left ventricle by an aperture at the base of the septum: the pulmonary artery was small and atrophic, it was not obstructed. there was no trace of the ductus arteriosus: the aorta passed back over the right bronchus and gave off four main branches. both subclavian arteries arose to the right of the right main bronchus and the left subclavian artery passed behind the trachea forming a "vascular ring"

Congenital deformities comprise about 2 per cent of patients afflicted with heart disease; but until February, 1945, when Alfred Blalock and Helen Taussig of Johns Hopkins Hospital reported their first two successful cases of Fallot's tetralogy treated

by subclavian pulmonary anastomosis, no surgeon had given the matter serious attention. This brilliant work not only proved that many "blue children" could be dramatically relieved of paralysing symptoms, but, by stimulating research all over the world it opened a new field of surgical endeavour.

A knowledge of the development of the heart is necessary in the management of these cases. The author recommends that the matter be studied in "Human Embryology," by



(From "Malformations of the Heart," by Thomas Peacock, 1858.)

FIG. 296. Lithograph of a specimen removed from a patient under the care of [illegible]

ary valves; the pulmonary arteries were hypoplastic, the left ventricle was small and the aorta was dextraposed, there was a ventricular septal defect.

Hamilton, Boyd, and Mossman. The principal milestones are these: at the third week of fœtal life the primitive vascular tube has already appeared: at first it traverses the pericardial cavity in a straight line in the long axis of the body. This blood tube, which is the anlage of the heart, grows more quickly than the pericardial sac, so that it doubles upon itself in the shape of the letter "S." The loop so formed becomes the ventricles and the bulbus cordis from which the aorta and the pulmonary arteries are derived. Meanwhile, at the caudal end of the pericardium, the future venous channels differentiate to

form the sinus venosus and the common atrial cavity, situated above and behind the ventricles. By the fourth week the beginnings of the heart chambers are apparent, and during the fifth week the septum primum and secundum have divided the *atrium** into right and left parts, and the whole of the sinus venosus has been absorbed into the right atrium. The valves develop at this time, but the interventricular septum does not grow until the eighth week.

Thus the heart appears and is completed in its permanent form between the twentieth and fiftieth days of fœtal life; but the foramen ovale and the ductus arteriosus remain open until birth. For this reason the extrinsic factors which are relevant to the development of congenital abnormalities operate during this brief period. These factors have only recently been investigated and knowledge is scanty, but the following may be relevant: virus diseases, such as German measles in the mother, vitamin deficiencies, deformities of the fœtal spine, syphilis, and fœtal endocarditis.

A child, born with a congenital deformity of the heart, may have other abnormalities such as arachnodactyly and mongolism.

A syndrome of some interest, described by Marfan, is not uncommon in congenital heart disease—in this the patient is tall and spare in build, has arachnodactyly, dislocation of the eye lens, and cystic medionecrosis of the aorta. This is thought to be a mesodermal dysphasia, and its importance is that the surgeon who operates on such a patient can expect to find the large arteries poor material for anastomosis.

As regards the heart itself it is usual to find one abnormality, or group of abnormalities, whilst the remainder of the organ develops as normally as possible considering the unusual circumstances imposed upon it. The clinical significance is that enlargements of this or that region of the heart can be important pointers towards accurate anatomical diagnosis, and it is unusual to find a part of the heart large unless it has been submitted to increasing strain.

It has been found that the majority of deformities associated with a large heart at birth are soon fatal, and the reason is that, at birth, the circulation of the infant is importantly changed. If the strain imposed by the deformity upon the right ventricle "in utero" has been such as to cause dilatation it is not likely that the child can live after birth. Whilst the fœtus is "in utero" its heart pumps the blood around the systemic circulation under conditions which are constant. At birth, not only is the pulmonary circulation added, but the heart must cope with varying demands of output. Before birth the brunt of the work falls upon the right ventricle; after birth it is the left which carries the chief load. The change over from one circulation to another is not immediate, and so it happens that some patients who have congenital deformities may, at first, appear to be normal. The foramen ovale and the ductus arteriosus may remain anatomically open, though they be physiologically closed, and in some congenital abnormalities the well-being of the infant can depend upon the ductus remaining patent. For instance a baby, having Fallot's tetralogy, may be well as long as blood can get from the aorta to the pulmonary artery by way of the ductus: such a child may die when the ductus closes.

Some patients who have congenital heart disease are blue (cyanosed) and some are not. The presence or absence of cyanosis depends upon the anatomical deformities in the heart itself. If cyanosis is present it may be constant or intermittent and is of the "central" type (q.v.) it is due to the fact that some venous blood is circulating in the systemic vessels and for this to be possible two things are necessary. First an abnormal communication must exist between the right and the left sides of the heart—i.e. a septal

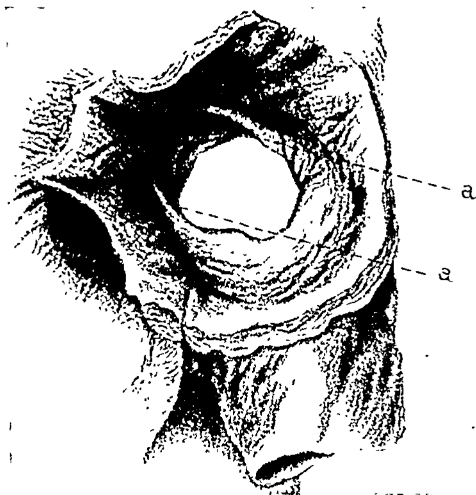


FIG. 297. Lithograph from Thomas Peacock's book on Malformations of the Heart (1858) It shows the foramen ovale in which the process of closure has never been completed, and the cornua of the valve are widely apart.

defect for example—and, secondly there must be a reason why the pressure on the right side is higher than that on the left—i.e. an obstruction in the pulmonary outflow tract—otherwise the venous blood would not flow into the systemic circulation.

The fundamental point which Blalock and Taussig were the first to emphasize was that in the majority of the cyanotic group of congenital deformities the principal defect from which the patient suffers is that not enough blood circulates through the lungs. Having this in mind Campbell and Paul Wood have recommended that it would be practical to divide the patients into those having an increased blood flow through the lungs and those whose lungs are starved.

form the sinus venosus and the common atrial cavity, situated above and behind the ventricles. By the fourth week the beginnings of the heart chambers are apparent, and during the fifth week the septum primum and secundum have divided the *atrium** into right and left parts, and the whole of the sinus venosus has been absorbed into the right atrium. The valves develop at this time, but the interventricular septum does not grow until the eighth week.

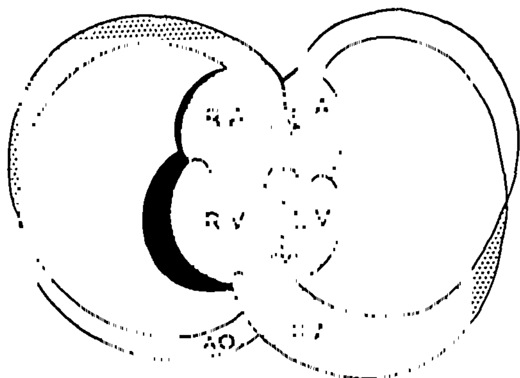
Thus the heart appears and is completed in its permanent form between the twentieth and fiftieth days of fœtal life; but the foramen ovale and the ductus arteriosus remain open until birth. For this reason the extrinsic factors which are relevant to the development of congenital abnormalities operate during this brief period. These factors have only recently been investigated and knowledge is scanty, but the following may be relevant: virus diseases, such as German measles in the mother, vitamin deficiencies, deformities of the fœtal spine, syphilis, and fœtal endocarditis.

A child, born with a congenital deformity of the heart, may have other abnormalities such as arachnodactyly and mongolism.

A syndrome of some interest, described by Marfan, is not uncommon in congenital heart disease—in this the patient is tall and spare in build, has arachnodactyly, dislocation of the eye lens, and cystic medionecrosis of the aorta. This is thought to be a mesodermal dysphasia, and its importance is that the surgeon who operates on such a patient can expect to find the large arteries poor material for anastomosis.

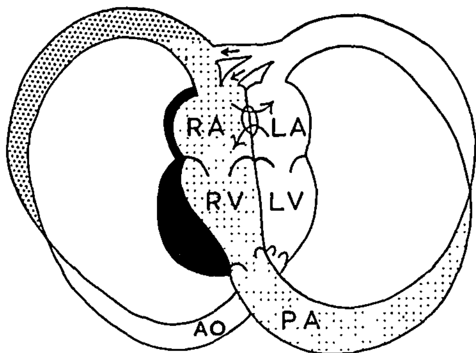
As regards the heart itself it is usual to find one abnormality, or group of abnormalities, whilst the remainder of the organ develops as normally as possible considering the unusual circumstances imposed upon it. The clinical significance is that enlargements of this or that region of the heart can be important pointers towards accurate anatomical diagnosis, and it is unusual to find a part of the heart large unless it has been submitted to increasing strain.

It has been found that the majority of deformities associated with a large heart at birth are soon fatal, and the reason is that, at birth, the circulation of the infant is importantly changed. If the strain imposed by the deformity upon the right ventricle "in utero" has been such as to cause dilatation it is not likely that the child can live after birth. Whilst the fœtus is "in utero" its heart pumps the blood around the systemic circulation under conditions which are constant. At birth, not only is the pulmonary circulation added, but the heart must cope with varying demands of output. Before birth the brunt of the work falls upon the right ventricle; after birth it is the left which carries the chief load. The change over from one circulation to another is not immediate, and so it happens that some patients who have congenital deformities may, at first, appear to be normal. The foramen ovale and the ductus arteriosus may remain anatomically open, though they be physiologically closed, and in some congenital abnormalities the well-being of the infant can depend upon the ductus remaining patent. For instance a baby, having Fallot's tetralogy, may be well as long as blood can get from the aorta to the pulmonary artery by way of the ductus: such a child may die when the ductus closes.



LATE STAGE. Right to left shunt (and mixed shunt) Right atrial and right ventricular hypertrophy Pulmonary artery enlargement.

FIG 299. Atrial septal defect. Right to left shunt.



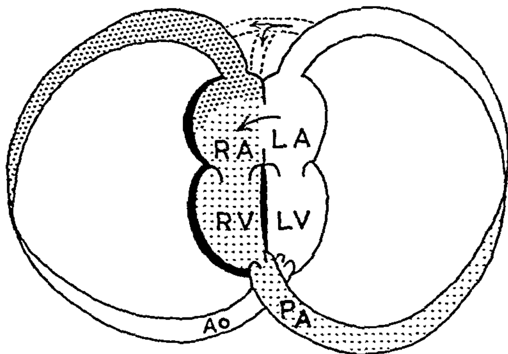
Veins draining into superior vena cava and/or left atrium Right atrial and right ventricular hypertrophy enlargement of pulmonary artery, and aortic hypoplasia NOTE An atrial septal defect may be present as an additional anomaly.

FIG 300. Atrial septal defect with anomalous veins.

ATRIAL SEPTAL DEFECTS

An *atrial septal defect* is an opening, which is not a valve, in the septum between the two atrial cavities. The remarks which follow concern those cases in which the defect is the chief or the only cardiac abnormality.

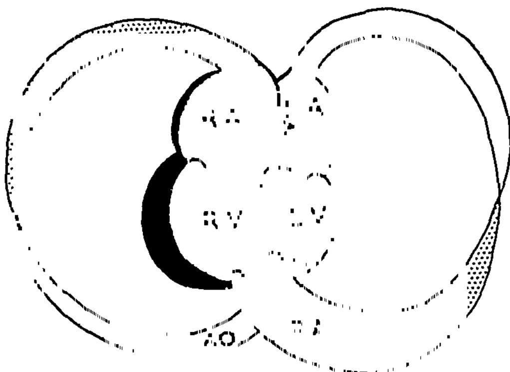
Surgical Anatomy. In the beginning there is only one atrial cavity which becomes imperfectly divided between the fourth and the fifth weeks, by the growth of the septum primum from the dorsal wall downwards and forwards. Arrest in the development of



EARLY STAGE. Left to right shunt (with temporary reversals).
NOTE. The mitral valve may be abnormal = Lutembacher's syndrome. There may be associated anomalous drainage of pulmonary veins.

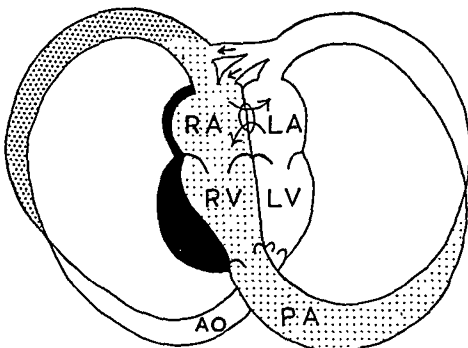
FIG. 298. Atrial septal defect. Left to right shunt.

this early partition leaves an opening between the two atrial cavities in the front and the lower part of the structure. The opening is called the *ostium primum*. If it persists it forms one type of atrial septal defect and is particularly difficult to close because the lower margin of the ostium is close to the atrioventricular valves and to the auriculoventricular node. Normally the ostium primum closes and a new aperture appears at the top of the septum primum. This is the *ostium secundum*, or the *foramen ovale*. The foramen ovale is soon covered by a second septum which grows down from the wall of the right atrium as an arched partition, which has a free crescentic lower margin. The two septa are approximated like the pages of a book but because the lower margin of the septum secundum is free and the upper part of the septum primum is deficient (foramen ovale) a valve-like chink remains between the two atrial cavities. If this hole persists the condition is called a *persistent foramen ovale* and is of no significance because the pressure in the left atrium, being higher than that in the right keeps the flap against the hole. But if the septum secundum does not grow a persistent atrial defect occurs and is the commonest variety of these anomalies. Such a defect is situated towards the top and the back of the adult septum.



LATE STAGE. Right to left shunt (and mixed shunt). Right atrial and right ventricular hypertrophy Pulmonary artery enlargement.

FIG 299. Atrial septal defect. Right to left shunt.



Veins draining into superior vena cava and/or left atrium Right atrial and right ventricular hypertrophy enlargement of pulmonary artery, and aortic hypoplasia NOTE An atrial septal defect may be present as an additional anomaly.

FIG 300 Atrial septal defect with anomalous veins.

In addition to the above clear-cut deficiencies, there are a number of complicated anomalies due to partial developments of one or both septa; and, at the moment it is not always possible, before operation, to know exactly what anatomy will be found when the septum is exposed.

Atrial septal defects accounted for 17·5 per cent of Paul Wood's series of 400 proved cases of congenital heart disease.

According to Bailey the following varieties of atrial defect can occur.

(1) *Uncomplicated*

A. Patent foramen ovale. No shunt.
B. Complete absence of septum. Shunt in both directions, i.e. mixing.
C. Persistent ostium primum. Left to right shunt.*
D. Persistent ostium secundum. Left to right shunt.
E. Localized single or multiple defects at any point in the septum. Left to right shunt.

(2) *Complicated*

A. Atrioventricularis communis. Predominant left to right shunt.
B. Patent foramen ovale, with:
 (i) Pulmonary stenosis. Right to left shunt.
 (ii) Mitral stenosis. Left to right shunt.
 (iii) Ebstein's malformation. Right to left shunt.
 (iv) Anomalous venous return from lungs. Right to left shunt.
C. Atrial septal defect, with:
 (i) Pulmonary stenosis. Right to left shunt.
 (ii) Mitral stenosis. Left to right shunt.
 (iii) Ebstein's malformation. Right to left shunt.
 (iv) Tricuspid atresia. Right to left shunt.
 (v) Transposition of great vessels. Mixing.

The remarks which follow concern persistent ostium secundum.

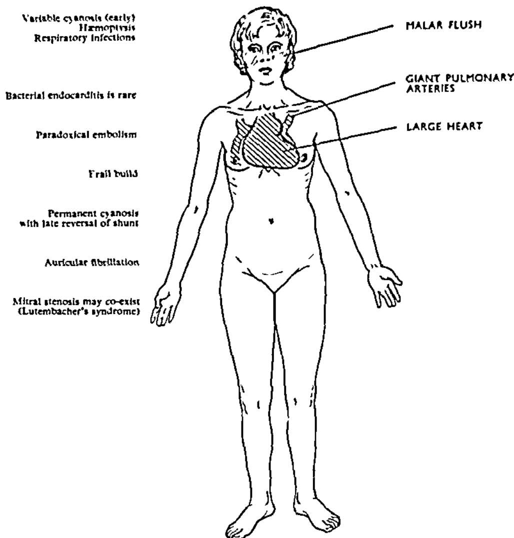
Diagnosis. It does not matter what the variety of the defect in the atrial septum (absence more or less complete, incomplete closure, or secondary perforation); what matters is the dynamic effect of the lesion.

The clinical picture depends on the fact that systemic blood flow is reduced, and pulmonary blood flow is increased; the discovered abnormalities all depend on this. With this defect there may be no clinical abnormality, and when there are any symptoms they are identical whatever the anatomy of the lesion.

With deviation of oxygenated blood from the left atrium through the atrial septum the aorta becomes hypoplastic and the pulmonary artery hypertrophied. As a result a low blood pressure with a small pulse pressure and pallor, and also peripheral cyanosis are common, because the systemic circulation is reduced and slowed. Late in the disease there is central cyanosis when the shunt is reversed. Intermittent cyanosis without heart failure is an important diagnostic point. Exertional dyspnœa is found, due to reduced systemic output, and to pulmonary plethora and stiffening. Bronchitis with hæmoptysis is common.

* In persistent ostium primum there is no septum below the defect and the cusp of the mitral valve is split so that the valve is incompetent. This complicated deformity requires special surgical treatment

The increased right heart and pulmonary blood flow result in right ventricular hypertrophy, with bulging of the lower part of the sternum if the hypertrophy occurs early in life, and with this a typical right ventricular cardiac impulse, felt best in the epigastrium. The pulmonary artery is enlarged, with percussion dullness in the second and third left



PROGNOSIS. 30–50 years (Bedford *et al*, 53 cases) Many years of semi-invalidism Operative risks—uncertain Operative results—can be excellent

FIG 301 Atrial septal defect

interspace and a visible and palpable impulse and often a systolic thrill. There is a pulmonary systolic murmur due to relative stenosis of the pulmonary valves and a loud and split second pulmonary sound, and in advanced pulmonary artery dilatation a pulmonary diastolic murmur due to incompetent valves. When the shunt is reversed and blood flows from right to left there may be an apical mid-diastolic murmur due to relative mitral stenosis, or in some cases to endocardial fibroelastosis or rheumatic endocarditis of the mitral valve. The E.C.G. assists in that right auricular hypertrophy may be discovered in tall P waves in leads I and II or a prolonged P.R. interval, or auricular fibrillation, flutter, or paroxysmal tachycardia (all evidence of atrial abnormality). In addition there is evidence of right ventricular strain and hypertrophy and eventually

right bundle branch block. X-ray examination shows a globular heart, a large pulmonary conus and main pulmonary artery, a large right atrium, and greatly enlarged pulmonary arterial branches with notable pulsations.

Rarely there is a tracheal tug due to an enlarged pulmonary artery and a paralysis of the left recurrent laryngeal nerve with hoarseness.

Natural History. It is twice as common in women. Rheumatic heart disease is often a complication with typical heart valve lesions and rheumatic vegetations on the rim of the septal defect. Subacute bacterial endocarditis is a rare complication, an interesting distinction between rheumatic heart disease with and without atrial septal defect. The bodily habit or build is often frail, and dwarfism, infantilism and delayed puberty are not rare, while mongolism and arachnodactyly are occasionally associated deformities.

The course is often long and without important symptoms or complication; and it is the commonest congenital heart disease to be found at postmortem in old age on persons dead of other disease.

Cyanosis may occur at times with long intervals of normality; in other cases upper respiratory infection and pulmonary infection are recurrent, cause hæmoptysis, and influence the course by impeding the pulmonary circulation and causing reversed shunt and cyanosis. It is likely that these pulmonary infections play a major part in advancing the disability. Thrombosis in the right atrium with pulmonary embolism and consequent pulmonary arterial hypertension is not rare. Because of the persistent right atrial stress right atrial fibrosis occurs with auricular fibrillation. Ultimately there may be atheromatosis of the pulmonary arteries and even aneurysm formation.

Pulmonary vein abnormalities are commonly associated with atrial septal defects and the inferior vena cava may open into the right atrium in such a position that blood from it easily enters the left atrium.

Investigations Necessary before Operation. If the presence of an atrial septal defect is suspected on clinical grounds, it becomes desirable to know its size and anatomical position, the presence of any other anomalies, and the hæmodynamics of the circulation between the two atrial cavities. To do this cardiac catheterization is essential. At first thought it might seem that these points could be quickly ascertained by taking samples of blood from the right atrial chamber and estimating its oxygen content. There are a number of fallacies in this: abnormalities of venous return to the atria may result in saturated blood being obtained in the right atrium; a patent foramen ovale may be present, and if the main stream of the blood in the right atrium happens to be directed towards the tricuspid valve an atrial defect can be missed. To circumvent these difficulties the following special tests should be done. oxygen saturations in the right atrium, the right ventricle and in the pulmonary artery, the pressures in the pulmonary artery; angiocardiography and perhaps the use of a ballooned cardiac catheter which can be engaged in the defect In some cases the cardiac catheter can actually be passed into anomalous veins

Indications for Surgical Treatment. Some patients who have an atrial septal defect need no active treatment. The pointers towards surgery are decreasing exercise tolerance, increasing cyanosis and evidence of increasing pulmonary congestion and right heart strain. It has been said that if the right ventricle is found on catheterization to be pumping more than 3 times more blood than the left ventricle the patient will die unless the circulation can be adjusted. In the future all cases will be corrected.

Surgical Closure of Atrial Septal Defects. Interest in the possibility of doing such an operation started with the work of Murray in 1945 onwards. In those days there was no method of opening the heart and operations were done blindly from outside. Murray showed that the gross anatomy of the heart was altered in these cases. When viewed from the right side of the chest the right atrium is voluminous and lies towards the front; it displaces the rest of the heart towards the left and the back. He devised a technique of

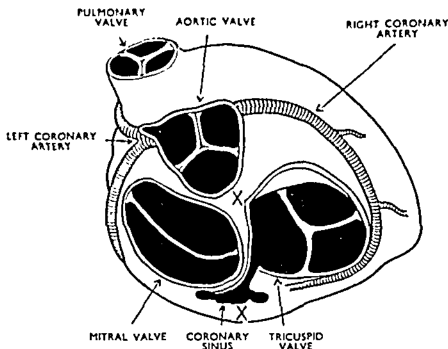


FIG. 302 Diagram showing the bases of the ventricles, exposed by removing the atria (after Gray's anatomy). The two crosses mark the points where the stitch, introduced to close an atrial septal defect, is fixed in Husfeldt's operation.

passing stitches across the common atrium and when he tied these stitches the redundant walls of the cavity were approximated in exactly the position where a septum should be.

Working on these lines other surgeons found it was possible to close some defects by invaginating one or both auricular appendages into the hole and some even filled the gap with bone or plastic buttons. These techniques are obsolete because they are inaccurate.

Bailey was one of the first to emphasize that before closing any defect a surgeon must at least put a finger into the atrial cavity, through the auricular appendage, in order to palpate the exact anatomy of the defect and also to find out the anatomical positions of the pulmonary veins, the coronary sinus and the valve of the inferior vena cava (the latter has, upon occasion, been mistaken for a septal defect with disastrous results). The mitral and tricuspid valves must also be explored. If the pulmonary veins open into the left side of the atrium, the surgeon can proceed to close the defect. If the pulmonary veins do not, then some special technique must be devised, for that particular case, which not only closes the defect but transposes the flow of oxygenated blood from the right to the left side of the atrial cavity. Bailey has worked out a number of relatively simple ways of achieving this end. In summary Bailey emphasized that not only must one close the

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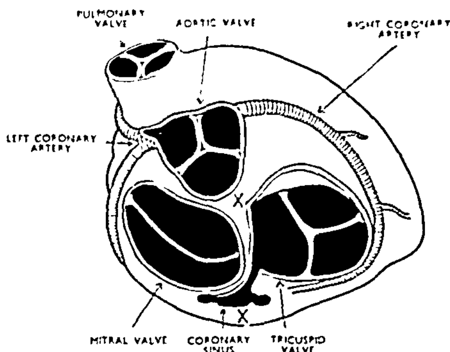


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VENTRICULAR SEPTAL DEFECTS

Pathological Anatomy. In the normal course of events the interventricular septum grows up from the apex of the heart, dividing the common primitive chamber into two parts and fusing with the septum in the bulbus cordis. If this process fails either totally, or in part, an abnormal communication persists between the ventricles. It has been customary to speak of these deformities as being of two types.



(*From "Malformations of the Heart" by Thomas Peacock, 1858.*)

FIG 303 Lithograph of a specimen showing a deficiency in the interventricular septum The deficiency is close to the aortic cusps, and this fact complicates the surgical treatment of many of the cases The specimen was exhibited at the Pathological Society of London by Dr. Quain in 1856, it was removed from a boy, aged 18, who had been cyanosed since the age of 2, and who died of cardiac failure.

The first was called "*La maladie de Roger*" (1897): in this there is a circumscribed hole in the membraneous part of the septum. The communication may be small or so situated that the shunt of blood from the left ventricle to the right is negligible, and there are no clinical disabilities. The heart is of normal size and shape, and the diagnosis is made by detecting a harsh systolic murmur and thrill to the left of the sternum in the third or fourth interspaces. If a large shunt is present serious disabilities occur and demand treatment.

Whether this shunt is from left to right or vice versa depends upon the relative pressures in the two sides of the heart; but in uncomplicated cases it is from left to right.

The foramen may be so large that the two ventricular chambers are virtually one. But 85 per cent of interventricular defects are about 15 mm. in diameter. The muscular type is generally not as serious as the membraneous which occurs higher, because the

defect, but at the conclusion of the operation the correct veins must discharge into the correct side of the heart.

Bailey's technique is to explore the atrial cavity with a finger inserted through the right auricular appendage. Being satisfied on the points described above, he keeps his finger in the atrial septal defect and sews a part of the redundant wall of the right atrium to the margins of the defect by invaginating the wall of the heart and creating a deep dimple on the surface. The blood from the superior vena cava can flow round the front or the back of this dimple without obstruction. This method is particularly suitable for closing holes in the top and posterior parts of the septum: it is not applicable to obliterating an ostium primum at the bottom of the septum, because the dimple may obstruct the tricuspid valve or the coronary sinus, the stitches may involve the auriculo-ventricular node or the bundle of Hiss, and the defect in the mitral valve is not corrected.

To overcome these difficulties Sondergaard, Husfeldt, Crafoord, and others have perfected another approach. In essence this consists of passing a stitch right through the heart in such a way that it travels down the septum and through the anterior margin of the defect. Prior to doing this the two vena cava have been mobilized and the whole of the back of the right atrium has been separated from the right pulmonary veins. The groove so developed is extended until the criss-cross muscle fibres which mark the posterior aspect of the atrial septum on the surface of the heart can be discerned. The two ends of the stitch are then tied in the groove and this obliterates the defect.

The tendency of work to-day is towards treating these defects by open heart operation, at which accurate suture of the hole can be done under direct vision. The first attempts on these lines were made by Gross who invented an "operating well" which he sewed to the wall of the right atrium. When the wall of the atrium was incised at the bottom of the well the blood in the heart rose up a short way (depending upon the intra-atrial pressure) into the well, and Gross was able to palpate the defect through the pool of blood, and to obliterate it with sutures or to sew in a piece of plastic sponge.

Recently the heart has been temporarily excluded from the circulation, either by using some type of heart lung machine or by cooling the patient; the blood in the atrium is then taken out and the defect seen and closed with stitches. This technique is effective and convincing. It is probable that, in the future, all cases will be done in this way.

Some patients who have an atrial septal defect also have mitral or tricuspid stenosis, and in these the abnormal valves can be opened in the usual way before closing the septal defect.

After closing an atrial defect the inter-atrial shunt is completely relieved and the strain upon the right heart and the lungs is relieved. The patient, from being desperately ill, can often be converted to a normal individual. But these operations have not yet reached the stage of perfection at which they can be done by all and sundry.

LUTEMBACHER'S SYNDROME

This deformity is due to the association of an atrial septal defect and congenital mitral stenosis; it is four times more common in women than men. The shunt of blood is from the left atrium back to the right; and the left atrium becomes aneurysmal. As soon as it is safe to close atrial septal defects, it will be possible to treat these patients, many of whom reach adult life, by overcoming the mitral obstruction and closing the septal defect. Both abnormalities can be corrected through the right atrium.

difficult and hazardous than those described for closing atrial septal defects. Kirklin has closed many ventricular septal defects using an extra-corporeal circulation. Excellent results have been obtained.

EISENMENGER'S COMPLEX

In this condition there is an interventricular septal defect, a displacement of the aorta to the right so that it overlies the right ventricle, and dilatation of the pulmonary arteries (aneurysmal enlargement is occasionally present). Pulmonary and aortic valve anomalies are common—bicuspid valves and cusps of irregular size. As a consequence of the constant overloading of the pulmonary circulation secondary changes develop in it with pulmonary hypertension and pulmonary atherosclerosis. Chronic respiratory infections and hæmoptysis are the main symptoms; and the physical findings are cyanosis increasing with the years, enlargement of the right heart and pulmonary arteries, a systolic murmur and thrill and often a diastolic murmur over the sternum.

Electrocardiography confirms right ventricular hypertrophy with strain and right bundle branch block.

Direct intracardiac measurement shows a very high right ventricular pressure and an equally high pulmonary artery pressure (which excludes pulmonary stenosis).

The prognosis is fair, some patients having reached old age.

The condition is mentioned for two reasons. First, it is always difficult to diagnose and mimic other lesions which would benefit from surgical intervention. Secondly, no operation of the Blalock or Potts type is of any avail because there is no pulmonary stenosis and because there is an adequate pulmonary blood flow and pulmonary hypertension. These patients are cyanosed because of the mixing of the venous and arterial blood in the heart itself, and the septal defect cannot easily be closed because the aorta is dextraposed. The usual description of Eisenmenger's complex states that the pulmonary artery is often aneurysmal in this condition; but increasing experience shows that when the artery is aneurysmal the diagnosis is more likely to be a patent ductus arteriosus with a reversed flow of blood into the aorta.

PULMONARY STENOSIS

Pulmonary stenosis may be pure, or complicated by over-riding of the aorta, and a ventricular septal defect (Fallot's tetralogy).

Surgical Anatomy of Pure Pulmonary Stenosis. Two anatomical varieties of obstruction occur

The first type is valvular and accounts for 80 per cent of all cases of pure pulmonary stenosis. The defect is limited to the pulmonary valve; the cusps are replaced by a diaphragm which projects into the base of the pulmonary artery like a nipple; at the top of the nipple there is a diminutive hole through which all the blood passes. Beyond the obstruction the beginning of the pulmonary artery is dilated and hypoplastic.

The second type is a defect in the primitive bulbus cordis and comprises 20 per cent of cases; it results in the development of an intracardiac chamber below the pulmonary valve The valve itself and the pulmonary artery are normal, and the deformity is situated in the infundibulum which is the anatomical name for the outflow part of the ventricle. The obstruction is usually localized and may be membraneous and diaphragmatic, or muscular, it may be situated immediately below the valve or about an inch or more away.

muscle in the septum contracts in systole and diminishes the size of the hole. The lesion may sometimes be complicated if, as a result of the defect, the conducting tissue is impaired or interrupted.

The deformity which interests the surgeon is that which occurs as a small foramen at the very top of the membraneous part of the septum; opening on the left side immediately underneath the aortic valve, and on the right in front of the median leaflet of the tricuspid valve. This may exist as an isolated defect or may be a part of such deformities as Fallot's tetralogy or Eisenmenger's complex. The remarks which follow refer to the cases in which the septal defect is the only abnormality present. It used to be thought that solitary defects of this kind could be ignored, but catheter studies and radiology have proved that in some cases the pulmonary circulation is constantly overloaded; that there is an undue strain on the right ventricle causing enlargement of the heart and an important left to right shunt. If the patient develops persistent pulmonary hypertension he is incurable surgically and dies.

The Diagnosis depends upon the typical systolic murmur and thrill which have been present from birth, in the third or fourth interspace to the left of the sternum, and finding these associated with a left to right shunt. The patient is not cyanosed, but the right ventricle is large and the pulmonary artery can become dilated.

The indication for surgical intervention is progressive limitation of exercise tolerance, heart failure, or rising pressure in the pulmonary circuit.

Treatment. At the present moment surgical treatment is tentative but successful operations have been performed. Bailey has fashioned a large pericardial flap which he has passed through the defect after locating it by direct palpation. Such a flap is not an ideal method of plugging the foramen because the pericardium undergoes degenerative changes when placed inside the heart and because the flap may obstruct the pulmonary outflow tract These operations are now obsolete.

Murray of Toronto contributed much to this subject and has operated upon a number of children successfully. He takes a long, straight needle, to which is attached a piece of thread and passes it, eye first, into the front of the heart at a point which he knows, from many dissections, to lie over the septum. He then introduces a finger into the ventricle and palpates the exact site of the defect and so guides the needle that it passes across the defect and out at the back of the heart. The thread is drawn through the heart and to the other end of the thread a strip of fascia lata is attached; this, in its turn, is pulled back across the defect. In this way several strips of fascia are introduced and when they are tied together and pulled taut the front of the myocardium is approximated to the back and the defect in the septum is closed. The mortality of this operation has been about 25 per cent. Murray has stressed the particular difficulties of closing these defects, which are almost always in the most inaccessible part of the heart; they involve the conducting tissue as well as the valve rings, and in some of the most serious cases there is only one ventricle In fact accurate closure demands open cardiotomy.

During 1955 Lillihei first treated a number of these cases by opening the ventricle and sewing up the hole in the septum under direct vision. To do this he diverted the patient's circulation from the heart and lungs and maintained life by using a cross circulation (q.v.) from a donor. The modern tendency is away from indirect or blind operations, and towards direct, open, surgery The technique has been applied not only to simple septal defects but to those forming part of Fallot's tetralogy. These operations are more

the pulmonary stenosis is so severe at birth that the only way in which the child can live is by reason of a right to left atrial shunt: it does not live long and is cyanosed from birth.

These remarks suggest that, as age advances, the lumen of the stenosis diminishes. It is more probable that as the child grows the muscle in the wall of the pulmonary outflow tract hypertrophies and the obstruction becomes more severe. In some mild



(From "Malformations of the Heart," by Thomas Peacock, 1858.)

FIG 305 Lithograph of a specimen from a patient who died at the age of 20 The heart shows a grossly hypertrophied right ventricle and stenosis of the pulmonary valves He also had a patent foramen ovale and, during life, was cyanosed on this account He suffered from urgent dyspnœa on exertion

cases there are no signs or symptoms. But the typical signs of pulmonary stenosis are a systolic murmur and thrill heard loudest in the pulmonary area, together with evidence of right-sided heart strain or failure The presence and degree of cyanosis and polycythæmia depends upon the interatrial shunt which may or may not be present. Squatting and right-sided aortic arch occur as commonly as in patients suffering from Fallot's tetralogy. Syncope on effort is relatively common. Preoperatively the diagnosis largely depends upon cardiac catheterization to measure the pressures in the right side of the heart and beyond the obstruction, and to detect the presence of an atrial septal defect; diagnosis also depends upon angiocardiography which shows that the systemic system

Depending upon the site of the infundibular obstruction there may or may not be a chamber between the valve above and obstruction below; the outer wall of this chamber may be muscular or membraneous.

Valvular and infundibular stenosis may coexist, and the latter may be due to muscular hypertrophy proximal to the valvular obstruction. The final anatomical diagnosis



(From "*Malformations of the Heart*," by Thomas Peacock, 1858)

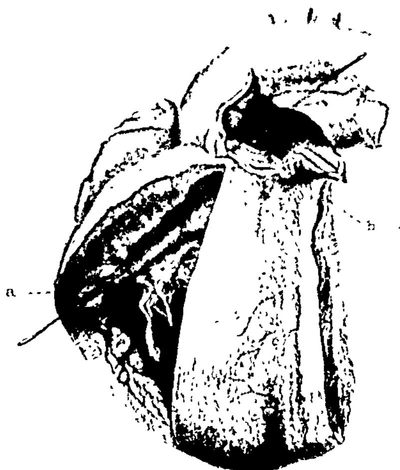
Fig. 304. Lithograph of a specimen showing the typical appearance of pulmonary valvular stenosis The normal cusps have been replaced by a nipple-like diaphragm; through which a fine probe has been passed into the pulmonary artery.

devolves upon the surgeon at operation, and the only way of doing it exactly is to examine the heart by intracardiac catheterization or open cardiotomy.

Clinical Features and Diagnosis. In the past the diagnosis of pulmonary stenosis has been overlooked or confused with that of Fallot's tetralogy. Some patients having mild obstruction live to old age and the defect is discovered at autopsy. In others the foramen ovale shuts in childhood; the pressure in the right ventricle rises, since all the circulating blood must then pass through the stenosis; and the patient is in danger of congestive heart failure. If the defect in the pulmonary outflow tract is severe and the foramen ovale patent, a right to left atrial shunt appears, and becomes more severe as the child grows: progressive cyanosis is then the outstanding sign, and the heart fails. In a few patients

the pulmonary stenosis is so severe at birth that the only way in which the child can live is by reason of a right to left atrial shunt. It does not live long and is cyanosed from birth.

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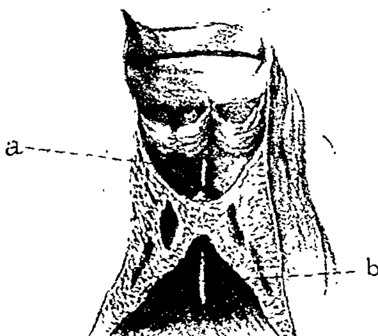
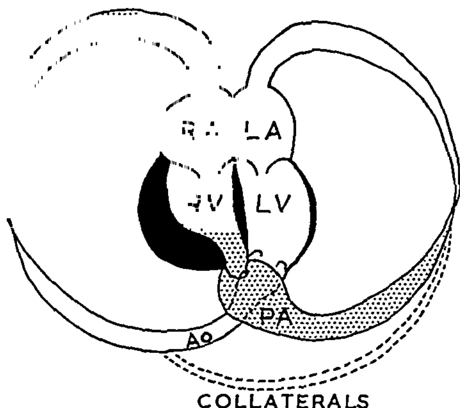


FIG. 306 Lithograph taken from the book, by Thomas Peacock, on Malformations of the Heart (1858). It depicts the pulmonary artery at the top, the pulmonary valves and an obstruction in the pulmonary outflow tract. The pulmonary valve cusps are only two in number, and one of them is imperfectly divided The obstruction lies in that part of the right ventricle known as the infundibulum.



Valvular and/or infundibular stenosis Post-stenotic dilatation of pulmonary artery Right ventricular hypertrophy NOTE. The post-stenotic dilatation of the pulmonary artery is a useful point of differentiation from Fallot's tetralogy in which the pulmonary artery is not large

FIG. 307. Circulation in pulmonary stenosis showing post-stenotic dilatation.

fills slowly from the lungs and not directly from the right ventricle Brock and Campbell state that the most important single radiological sign of pulmonary valvular stenosis is post-stenotic dilatation of the pulmonary artery.

Prognosis. Considering the wide range in the degree of obstruction which can exist from patient to patient it is manifest that the prognosis will vary accordingly. Right heart failure is an important risk. Most patients die young—the average span in Abbott's series was 20 years—and few reached old age: thus, if operation is contemplated it should be done in early life. Brock has stressed the risk of sudden death both during everyday life and particularly at operation. He believes this to be due to coronary insufficiency and points out that if the condition of the patient suddenly deteriorates during operation the surgeon has only one possible remedy—to divide or remove the obstruction as quickly as possible.

Treatment. The first man to attempt to operate upon these patients was Doyen in 1913: he did not succeed. The surgical treatment of valvular and of infundibular stenosis is different in regard to the details of how the obstruction is to be overcome, but identical in that only by operating upon the obstruction itself can the condition of the patient be ameliorated. It follows from what has been said already that the surgeon must have an exact diagnosis, and if this is not available preoperatively he must examine the heart before deciding upon surgical treatment. In pure valvular stenosis the sinuses of valsalva are usually large or aneurysmal; the pressure in the pulmonary artery, which is dilated, is usually low or normal; the jet of blood coming through the pinhole aperture in the diaphragmatic valve can be palpated. By contrast, in infundibular stenosis the root of the pulmonary artery is not dilated and the infundibular chamber may be visible. In such cases exact diagnosis is made by incising the right ventricle towards the apex of the heart and passing a sound, or the finger, into the interior. In this way the obstruction can be palpated and its anatomical site demonstrated. If doubt remains a catheter, connected to a pressure recording apparatus can be passed into the right ventricle. As the tip of the catheter travels from the ventricle through the narrow orifice of the obstruction the pressure falls, in this way not only the site of the block, but its length may be ascertained.

The Treatment of Valvular Stenosis

If the obstruction is found to be in the valve itself, the procedure is as follows. The operation described is that devised by Brock who has done most of the pioneer work. The first man to do a pulmonary valvotomy successfully was Sellors.

The heart is exposed through a long antero-lateral intercostal incision. The pericardium is incised parallel to and in front of the phrenic nerve. If the right ventricle does not present adequately, the heart can be rotated slightly by exerting traction upon the anterior flap of the pericardium An incision is made into, but not right through, the wall of the ventricle. The chamber of the right ventricle is entered by passing in a small metal sound (rather like a simple Clutton's urethral sound) and, using this as a feeler the anatomy is determined. If doubt remains the pressure in the right ventricle and the pulmonary are recorded electromanometrically. There is no need to insert a number of mattress, or stay, sutures in the myocardium on either side of the proposed incision because hæmorrhage can be controlled by digital pressure with the pulp of the finger. To overcome the obstruction a special curved, flat, knife is passed into the heart and through the stenosed orifice of the valve. This knife makes two lateral incisions in the membraneous

part of the valve and these are enlarged by a series of special dilators. The object of the manœuvre is not only to increase the lumen but to leave two flaps which can function, and prevent regurgitation. When the instruments have been withdrawn the incision in the myocardium is closed with two or three mattress stitches. The pericardium should be loosely closed to avoid tamponade and the pleural cavity drained for 24 hours.

Some surgeons prefer to do the operation by approaching the valve from the pulmonary artery. Husfeldt, for instance, has devised a slender guarded knife which he inserts into the pulmonary artery and juggles through the narrow orifice of the valve into the right ventricle. By means of a screw on the handle, the knife blades are laid bare and the cutting instrument is drawn through the valve from below upwards.

None of the operations described above is done under direct vision and all are agreed that, to cut or rupture the stenosed valve blindly, is apt to lead to poor results. Many surgeons have treated these cases by cooling the patient, clamping the cavæ, the aorta, and the pulmonary artery, opening the pulmonary artery deliberately, and cutting the valve under direct vision. The results of operations of this type have been very satisfactory in that a definite fall of pressure in the right ventricle is always achieved.

The Treatment of Infundibular Pulmonary Obstruction

This was first successfully achieved by Brock. He pointed out that the obstruction was generally limited, and he devised a cold punch which he inserted into the ventricle below the obstruction. The isthmus of obstructing tissue was engaged in the cutting part of the instrument and the excised fragments passed into the interior of the punch which was hollow. After punching a fairway through the obstruction the lumen was perhaps enlarged by graded dilators. In subsequent operations this technique was modified. If the infundibular chamber was large, and its wall muscular, the punch was inserted distal to the obstruction. This had the advantage that, if a valvular stenosis was present as well as one in the infundibulum, both anomalies could be dealt with through the same incision.

Bailey and others have also shown that if a punch cannot, for some reason, be engaged in the obstructing isthmus fairly good results can be achieved by digital dilatation.

The mortality of these operations varied at first between 20 and 30 per cent, but the results, which were excellent in the early follow-ups, may not be good permanently, for there is evidence that in some of these cases the whole of the pulmonary outflow tract is abnormal.

Since these early operations many surgeons have devised techniques for removing infundibular stenosis by inserting a punch from the pulmonary artery in a retrograde fashion: all these attempts are influenced by the fact that operations done through the pulmonary artery are not so likely to result in ventricular fibrillation as those done through the ventricles.

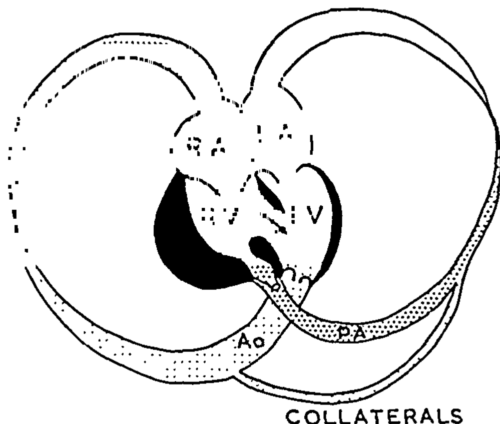
The present tendency is to do a direct intracardiac operation whenever possible—using either "cooling," some form of heart-lung machine, or a cross circulation. It is only in this way that all the obstructing tissue can be totally and accurately removed

FALLOT'S TETRALOGY

Fallot's tetralogy is the name given to a combination of four congenital cardiac deformities of which pulmonary stenosis is one. The others are, a patent interventricular septum, over-riding of the aortic root, and hypertrophy of the right ventricle. These

defects have two principal results: blood from the right ventricle mixes with that from the left, and the lungs are starved of blood. The clinical result is to produce "central" cyanosis and to limit exercise.

Fallot's tetralogy accounts for 66 per cent (Paul Wood) of all infants suffering from congenital cyanotic heart disease, and is the commonest cause of congenital heart disease in children who are born blue and who survive infancy.



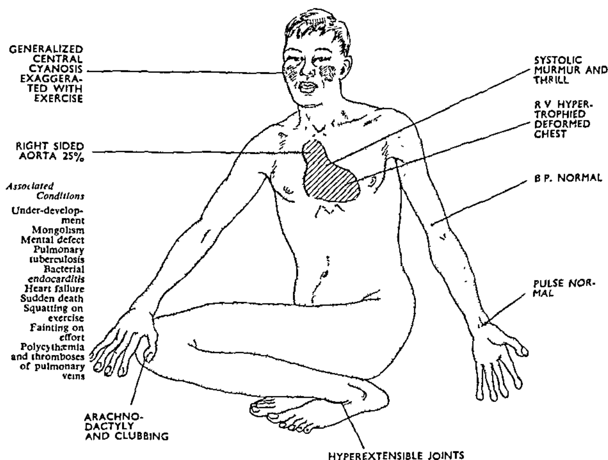
Valvular and/or infundibular stenosis Dextroposed aorta Right ventricular hypertrophy. Interventricular septal defect NOTE The development of collateral circulation varies greatly in individual cases.

FIG 308 Circulation in Fallot's tetralogy

Surgical Anatomy. Fallot's tetralogy is due to failure of rotation of the bulbus cordis, associated with an anomaly of the interventricular septum and a deformity of the pulmonary outflow tract. Externally the heart may be of normal appearance in mild cases, but the right ventricle becomes hypertrophied and the typical shape has been described as resembling a French sabot. If the stenosis is valvular the pulmonary artery is large and the sinuses of Valsalva are dilated; the normal valves are replaced by a perforated conical diaphragm in the origin of the pulmonary artery. If the stenosis is infundibular or muscular the pulmonary artery looks normal, the valve is normal but the outflow tract is obstructed; the subvalvular chamber may or may not be visible upon the surface of the heart. The lungs are starved and the systemic vessels contain venous blood.

Fallot's tetralogy is associated with a right sided aortic arch in 25 per cent of patients; and other congenital deformities such as arachnodactyly, webbing of the fingers and toes are common, but it is unusual for these associated deformities to be of decisive importance in treatment.

In many patients the body attempts to overcome pulmonary ischæmia by developing an extensive systemic collateral supply to the lungs. The vessels concerned arise from the bronchial arteries, the intercostals, the phrenics and the œsophageal vessels. They can form such a network of new channels as to produce murmurs, and can be demonstrated on radiographs. The new vessels carry systemic blood to the lungs, where it is oxygenated and returned to the left side of the heart. In some patients suffering from severe pulmonary



PROGNOSIS Invalidism with much family upset 15–20 years Operative risks—10 per cent. Operative results—greatly improved exercise tolerance, improved growth, but other risks remain, i e bacterial endocarditis, heart failure

FIG 309. Fallot's tetralogy.

ischæmia no collateral vessels exist: the presence or absence of these vessels bears no relation to the degree of pulmonary ischæmia from which the patient suffers, and it is not known why they grow in some cases and not in others.

Diagnosis. The children are generally of normal intelligence, but may be physically underdeveloped and have often been treated in their homes as invalids and liabilities. The typical signs and symptoms occur in a child who has been cyanosed from birth, but if the ductus arteriosus has remained patent, cyanosis may not be obvious until it closes. There is "central" cyanosis, polycythæmia, clubbing of the fingers, limitation of exercise tolerance and a tendency to squat, all of which become more and more obvious as the patient grows older. Dyspnœa on exercise is the most important symptom. The murmurs are not pathogonomonic, but in most cases a systolic murmur can be heard in the third or fourth left interspace to the left of the sternum. A thrill may be palpable.

The radiographs of the lungs show a diminution of blood vessels, and there is a depression between the aortic knuckle and the ventricles because the pulmonary arteries are hypoplastic. Cardiac catheterization with angiocardiography is generally necessary to clinch the diagnosis. The systolic blood pressure in the right ventricle approximates to

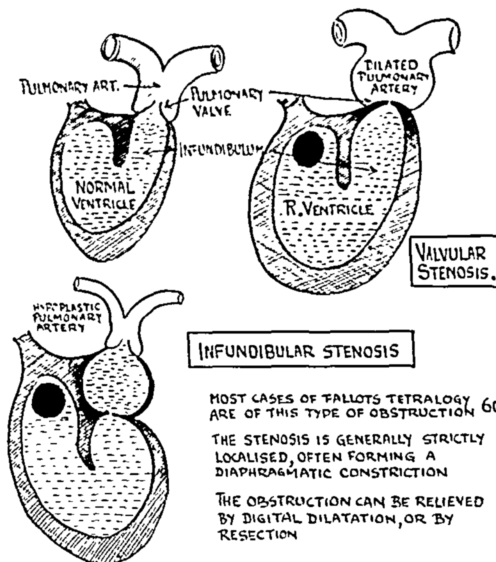


FIG. 310

the systemic systolic pressure, and if the tip of the catheter can be passed into the pulmonary artery the pressure drops suddenly, to less than a mean of 10 mm. of mercury, as it penetrates beyond the obstruction. The low oxygen saturation of the systemic blood proves that a veno-arterial shunt is present.

Prognosis without Surgical Treatment. Campbell, who has analysed a series of 340 patients suffering from cyanotic heart disease stated that only 1 child in 2 is likely to survive until the age of 7; 1 in 4 may reach 14 years; and less than 1 in 10 will survive till 21.

The common cause of death was asphyxia.

Not all patients suffering from Fallot's tetralogy are severely disabled, nor is the severity of the cyanosis a guide to the need for surgical treatment. The best clinical

method of assessing the disability is to apply simple exercise tests to note the increase in cyanosis and to measure how long it takes the pulse rate to return to its resting level. The safest time to advise surgical treatment is between the ages of 5 and 10, but progressive disability calls for earlier operation.

Treatment. Blalock and Taussig were the first (1945) to show that dramatic results could be achieved in many of these patients, and Blalock has now operated upon more than 1,000 children. In the operation he devised, one subclavian artery was anastomosed end to side to a pulmonary artery or to one of its branches. Usually the right subclavian was preferred because it aligned more easily than the left which was apt to be flattened like a ribbon if turned down across the arch of the aorta. In some cases, where the pulmonary artery was very small, it could with advantage be anastomosed to the subclavian by dividing it across and joining the two vessels end to end. Before any of these operations could be done the surgeon must know the anatomy of the branches of the aorta and of the aortic arch itself; he must have demonstrated that the patient has pulmonary arteries, and he must be sure of diagnosis.

Blalock's operation carries a mortality of about 10 per cent in the hands of the expert, but the risk is far higher for those surgeons who are not constantly doing this type of work. The results are often incredibly good, for not only are the polycythæmia, cyanosis, and squatting overcome but the exercise tolerance is dramatically increased, and the child can often lead a normal life. As yet the operations have not been done long enough to know whether the ultimate risks are the same as those associated with a naturally occurring patent ductus arteriosus: but, to date, this does not seem to be the case.

In 1946 *Potts of Chicago introduced a modification of Blalock's principle.* By devising a special clamp which isolated a small segment of the wall of the aorta without interrupting the main flow of blood through the vessel, Potts was able to anastomose the left pulmonary artery directly to the concave margin of the aortic arch. This operation achieved its results in precisely the same way as that devised by Blalock. Most surgeons prefer Blalock's operation, if the anatomy is suitable for either procedure, because it is difficult in Potts' operation to make an anastomosis of the correct size (i.e. $\frac{5}{16}$ in.). If the anastomosis be too large the patient develops pulmonary œdema and if too small thrombosis and obliteration occur. On the other hand Potts' operation is easier to do than Blalock's in children under 3 years of age.

Technique of Blalock's Operation

Blalock's operation is done under general anæsthesia in which the important point is that adequate ventilation with plenty of oxygen is continuously maintained. A transfusion is set up, but blood is not given, unless bleeding occurs, because the patient already has polycythæmia. The drip may be kept patent with dextran or saline solution.

Most British surgeons open the chest on the side of the arch of the aorta and there are two reasons for this: it is technically easier to use the left subclavian artery for the anastomosis than the right, and if the surgeon, having examined the anatomy, elects to do Potts' operation in preference to Blalock's, this can be done without altering the incision. A postero-lateral intercostal incision gives good access to the superior mediastinum. In some cases the pleural cavity will be found obliterated by vascular adhesions and these can present a formidable problem at the outset, because the adhesions carry an important collateral blood supply to the lungs. Such adhesions have to be divided carefully and

hæmostasis secured before one can proceed with the operation Having reached the mediastinum, the next problem is to establish the diagnosis and, as Brock has repeatedly stressed, this can only be done by opening the pericardium and examining the pulmonary outflow tract, in the ways described above. At the moment the pericardium is opened these patients may develop cardiac arrest or dangerous arrhythmias if these occur the pericardium should be temporarily closed and the appropriate measures applied. If the surgeon elects to do a Blalock operation the pericardium is closed with interrupted stitches and the next step is to deal with a multitude of engorged collateral vessels which enter the hilum of the lung from the internal mammaries, the phrenic and the mediastinal arteries. These vessels run in the loose areolar tissue immediately deep the mediastinal pleura and if the pleura is incised in one straight line from the apex of the chest to the front of the hilum the flaps can be turned forward and back taking the collaterals away from the pulmonary and subclavian arteries. The pulmonary artery is dissected clean from its origin to its insertion into the lung, and in doing this a ductus arteriosus may be found and should be preserved if it is patent. When the pulmonary artery has been cleaned it is well to apply a Blalock's clamp for a few minutes in order to test the effect of diverting the whole blood supply to one lung: usually there is no alteration in the condition as a result of this manœuvre, but if the circulation deteriorates or the heart becomes irritable the surgeon must wait, allowing the lungs to be fully ventilated for a time. The subclavian artery is now dissected out from its origin to the dome of the chest. In so doing anomalous branches may be encountered and must be tied off. At the top of the mediastinum the first branches of the subclavian artery come off, and if the vertebral arises low the dissection must be carried beyond this branch, so that a long enough piece of subclavian may be available to reach the pulmonary artery. The subclavian artery and its branches must be securely tied and a bulldog clamp applied to its origin near the aorta. When this clamp has been tested the subclavian artery is divided at the preselected spot and in such a way that the open end is slightly funnel shaped. The adventitia is then meticulously cut away from the artery for, if this is not done, it gets entangled in the stitches used for the anastomosis. The Blalock's clamps are then applied at either end of the exposed part of the pulmonary artery and a transverse incision is made into this vessel between the clamps. This incision should correspond in length to the size of the end of the cut subclavian. The anastomosis is done using 00000 black silk on a small, round-bodied needle. The stitch is continuous and everting. There is no room for error in this anastomosis; if a stitch is passed wrongly or tears out, it creates a hole in the vessel which leaks, every unnecessary hole may have to be cobbled up and this narrows the lumen When the anastomosis has been completed both the Blalock clamps should be taken off at the same time and no bleeding should occur: it is much better to take off both clamps together than to take one at a time. If persistent bleeding occurs the clamps must be reapplied and gentle pressure tried before additional stitches are put in. In most cases this suffices, but sometimes the suture line needs reinforcement. Having secured a dry field the surgeon can test the success or otherwise of his operation, because there should be a continuous machinery thrill in the pulmonary artery. The colour of the patient and the peripheral circulation do not always improve straight away because of the polycythæmia. The chest should be closed in the usual way and temporary pleural drainage provided. Oxygen therapy may be needed after the operation, but heparin is not necessary.

If the subclavian artery is not sufficiently long to reach the pulmonary artery without tension the gap may be bridged by using an arterial graft, joined to the subclavian at one end and to the pulmonary artery at the other.

The operation described above is that originally devised by Blalock and his associates. It has found much favour wherever it has been practised; but it is certain that, in the future, it will be displaced by other procedures which aim at diminishing the number of deformities, rather than increasing them. Lillehei, Kirklin and other surgeons, using various types of extra-corporeal circulation, have opened the right ventricle in these children, overcome the obstruction in the pulmonary outflow tract and then closed the interventricular septal defect. Such an operation makes the heart and the circulation normal. The defects of Blalock's operation also apply to that described by Potts (q.v.): both operations will soon be obsolete

The Technique of Potts' Operation

The mediastinum is exposed in the same way as has been described for Blalock's operation. The aorta in the vicinity of the left subclavian artery is mobilized to such an extent that the special clamp devised by Potts can be applied. This clamp pinches off a small part of the aorta, where the anastomosis is to be made, but leaves the main channel free from obstruction. The left pulmonary artery is then exposed and a suitable segment isolated between two Blalock's clamps. The anastomosis should be about 4 mm. in length in a child, and made on the concave side of the aorta at the point where the vessel is crossed by the left pulmonary artery. One layer of finest black silk stitches, mounted upon a small, curved, needle is used. The anastomosis is covered by suturing the mediastinal pleura over it.

In some cases enlargement of the heart has been noted after anastomosis. This has generally happened within the first 2 months and was possibly due to the fact that the heart grows because it has more work to do and because its blood supply improves; it does not necessarily mean that pathological dilatation has occurred.

Pulmonary Valvotomy for Fallot's Tetralogy

Brock advocated another way of alleviating Fallot's tetralogy: he operates upon the obstructed pulmonary outflow tract directly, and does a valvotomy or an infundibular resection. He admits the success, and the relative safety, of Blalock's operation but points to these objections: it is an indirect attack on the problem and one which adds a fifth deformity to the four already present; moreover unrelieved pulmonary artery obstruction may be progressive But the problem of how to relieve the obstruction itself is not the same in Fallot's tetralogy as it is in pure pulmonary stenosis, because in the former the obstruction is more often infundibular (60 per cent) than valvular (40 per cent). Thus infundibular resection is necessary more often than valvotomy, and this operation carries a greater risk. Moreover in some cases the infundibular stenosis is not a simple stricture but a muscular deformity of the whole outflow tract; and finally it has been argued that even if success is achieved the lesion has merely been converted from one of Fallot's tetralogy to one of Eisenmenger's complex. To cure the patient the ventricular septal defect must be closed as well as relieving the pulmonary stenosis. This is the operation of the future.

In spite of these criticisms Brock has claimed good success with direct operations,

which he does in about 50 per cent of cases. He always explores the open pericardium and diagnoses the exact nature of the pulmonary obstruction before deciding which type of operation to do. Direct operations upon patients suffering from simple or complicated congenital pulmonary stenosis are feasible, they have a low mortality and the results are at least as good as those following a Blalock or Potts operation.

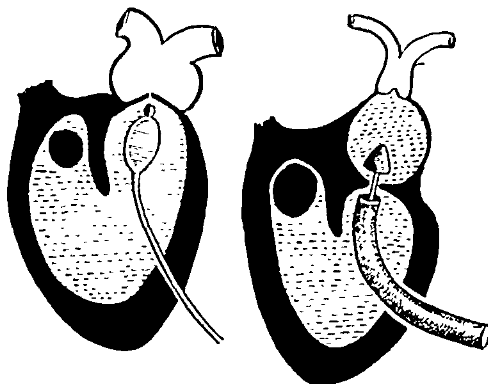


FIG 311

PLEURODESIS FOR FALLOT'S TETRALOGY

A third principle of treatment was introduced by Barrett and Daley (1949) who advocated pleurodesis to make the lungs adherent to the chest wall and the mediastinum, and so to encourage the growth of an adventitious blood supply from the systemic arteries in the parietes. This manœuvre depends upon the observation that a patient can live, in the absence of pulmonary arteries, provided the lungs receive an adequate blood supply from systemic vessels. In 200 patients suffering from morbus ceruleus Brock stated that no anastomosis was possible in 11, i.e. 5·5 per cent.

The operation is simpler, and consequently safer, than those described above, but it does not succeed so dramatically. It is consequently reserved for patients who are too ill to stand a long procedure or so mildly affected that it may not seem justifiable to do an operation which carries a higher mortality. For the operation to succeed the patient must not develop a hæmothorax during convalescence.

The Complications after Operation for the Relief of Fallot's Tetralogy. One of the most surprising things in the whole field of surgery is how well most of these blue children tolerate major procedures upon the heart itself and the great vessels. The mortality of all the operations discussed above varies not only with the severity of the abnormality but with the skill and experience of the surgeon. Every surgeon who has worked in this field

has stated that many of the deaths which occurred in the first 100 cases were due to lack of technical skill, and that the only way to acquire this skill is to practise vascular anastomoses on animals or to work at first with a master.

Campbell and Deuchar reviewed the early and late results in 200 patients suffering from morbus cæruleus and treated by Blalock's operation.

Most patients who left the table in good condition needed oxygen for about 24 hours but, after that, convalescence was smooth. The immediate complications were hæmorrhage from the suture line, cardiac failure due to sudden changes imposed upon the circulation, cerebral and peripheral thrombosis, due perhaps to the polycythæmia, pulmonary embolism and pulmonary œdema. Transfusions were avoided unless there was definite evidence of bleeding.

In most cases the children were convalescent within a week and ready to leave hospital shortly after that.

If a good result has been achieved, the life of the patient and that of his family will often be transformed; some children will need special education and help before they can take their rightful place in society. There is no evidence to suggest that patients suffering from morbus cæruleus are mentally backward, but they have practically always been denied the life and teaching which a normal child gets.

Shortly after a successful operation it is usual for the heart to increase a little in size. This increase is not necessarily associated with adverse symptoms, and tends to stop after a few months. In some few cases it is a warning of impending cardiac failure.

Late Results of Operation In successful cases the colour of the patient is transformed, the exercise tolerance rises, the oxygen saturation of the blood rises, whilst clubbing and polycythæmia are diminished. The results achieved by various operations, and *without open cardiotomy*, are as follows (Brock, Sellors, Hill).

	Patients
Very good	99
Good	37
Improved	17
No improvement	6
No anastomosis possible	11
Deaths at or shortly after operation	24
Died at some later date	6

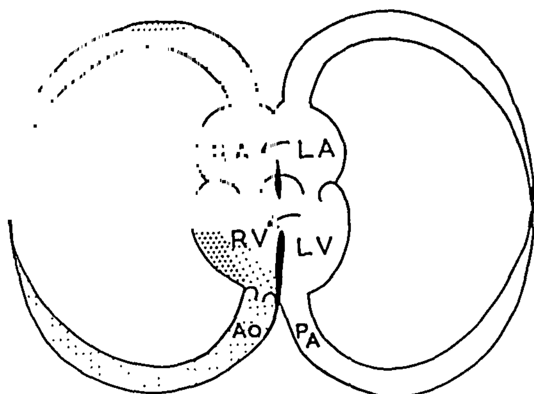
The majority of these patients had Fallot's tetralogy and many were seriously ill; in this group the overall mortality was 8 per cent and 75 per cent have been much improved. The patients classed as very good are well enough to lead practically normal lives. In patients having anomalies other than Fallot's tetralogy the mortality was more than 30 per cent and only 35 per cent were benefited.

The patients are generally much better soon after operation if the ultimate result is going to be favourable; the late annual mortality has not exceeded 1 per cent to date. In practically every successful case a continuous murmur can be heard at the site of the anastomosis, and if the patient has been improved and has not got a continuous murmur the reason is that new collateral vessels have formed as a result of the anastomosis. In such cases the anastomosis is almost certainly not patent.

In this English series neither bacterial carditis nor arteritis was a problem, but five patients developed cerebral abscess months after operation.

TRANSPOSITION OF THE GREAT VESSELS

This condition, which occurs almost as commonly as Fallot's tetralogy, remains an unsolved surgical problem. Untreated the prognosis is hopeless, but the heart and the great vessels, though abnormally arranged, would be capable of normal function if the deformities could be overcome. The condition is a challenge to surgery.



Aorta arising from right ventricle. Pulmonary artery arising from left ventricle. Atrial and/or ventricular septal defect. NOTE. A septal defect is essential for survival.

FIG. 312. Circulation in transposition of the great vessels.

The pulmonary and the systemic circulations are virtually separate, and not in continuity as they should normally be. The aorta arises anteriorly from the front of the right ventricle and the cavæ return as usual to the right atrium; the pulmonary artery arises posteriorly from the left ventricle and the pulmonary veins return to the left atrium. Life is only possible if a septal defect is also present and a patent foramen ovale is commonly found. In addition the ductus arteriosus is usually patent and the patient generally dies when this channel closes.

The circulation is complicated both before and after birth. Basically there is a good flow of oxygenated blood from the left ventricle to the lungs; and a poor flow of venous blood from the right heart to the systemic vessels. The left ventricle pumps most of the arterial blood from the lungs back through the lungs. When the pressure in this circuit rises, some arterial blood flows from the pulmonary artery to the descending aorta by way of the ductus. By contrast the right ventricle pumps its venous blood through the aorta to the head and extremities and it receives blood back from the cavæ, plus any additional blood which has gone through the ductus. It follows that with each heart beat more

blood returns to the right ventricle than it has pumped. Thus the pressure in the right atrium rises, the pressure in the left atrium falls; and the foramen ovale opens so that some mixing of the two independent circulations occurs.

Diagnosis. These babies are born blue and cyanosis is often intense. The pulses are equal in all the limbs, but cyanosis is most marked in the upper part of the body, and the abdomen. The children are feeble and dyspnœic. 25 per cent die in the first month of life and 75 per cent in the first 12 months. The heart, which is small at first, enlarges rapidly. The lungs are congested. Radiologically the heart shadow is narrow at the base and broad over the ventricles. The E.C.G. shows right axis deviation. Murmurs are present but are not of help in the diagnosis.

Treatment. The ideal management of such a case would be to divide the aorta and the pulmonary arteries and to carry out a cross anastomosis between these two vessels. The difficulties are great for not only are the infants frail and feeble but the circulation must be totally interrupted for a considerable time; and, most important, the origins of the coronary arteries must originate from the stream of oxygenated blood. This operation has been attempted and some success claimed. Less drastic measures consist in transposing all or some of the vena cavæ and the pulmonary veins; or in creating a permanent atrial defect and then anastomosing the subclavian artery to the pulmonary artery.

RHEUMATIC HEART DISEASE

Introduction

Mitral stenosis was regarded as a possible surgical problem by Sir Lauder Brunton in 1902; but it was not until 20 years later that the first patients were operated upon by Cutler and Beck in America, and by Sir Henry Souttar in England. The latter devised and practised digital fracture of the stenosed mitral valve, and he performed the operation through the auricular appendage using virtually the same technique as is followed today. He was unable to proceed with the work at the time because he lacked the support of his medical colleagues. Between 1923 and 1945 the matter lay dormant and it was not until after the second World War that progress began again.

Many different surgeons have contributed to advances in this subject since 1945, but Harken of Boston, Bailey of Philadelphia, Brock and Sellors of London, Logan of Edinburgh, Dubost and Mathey of Paris are pre-eminent. The surgery of rheumatic heart disease has so far been confined to the treatment of valvular disorders.

The nature of rheumatic pancarditis, which initiates the destruction of the valves, remains in doubt, but the evidence suggests that it is not an infective process. The acute disease particularly affects poor children living in damp surroundings, and its incidence runs parallel with tonsillitis due to hæmolytic streptococci. There is an hereditary disposition and an association with Sydenham's chorea, subcutaneous nodules, and erythema marginatum. The last is associated with rheumatic fever in 15 per cent of cases. Some 50 per cent of patients give no history of tonsillitis, acute rheumatism, or of chorea.

It is generally believed that the "rheumatic state" is an allergic phenomenon produced in a patient who is sensitive to the products of hæmolytic streptococci. There is no evidence that the changes which destroy the valves are due to bacterial infection. If patients who have suffered from rheumatic fever recover, the pancarditis and polyarthritis generally settle and some have no further trouble, but such an attack in childhood

will almost certainly leave its effects. In a small number the heart will be grossly damaged and death will occur after a few months. In others after a period of 2-25 or more years, during which the patient is symptomless, symptoms will appear. A simple formula will serve to illustrate the course after rheumatic fever: 10 years without symptoms, 10 years with slowly progressive symptoms, and then 10 years of recurrent heart failure.

The surgeon can intervene in this pathological process to remedy the mechanical disabilities. To do this effectively he must know whether valvular disease is part of a continuous rheumatic process or whether, in the absence of specific signs of infection, it can be assumed that the rheumatic infection has ceased to act, and the deformities which remain are impaired muscle action and mechanical faults. If rheumatic pancarditis has never subsided operations to overcome obstruction may fail because muscle disease may predominate; but if progressive symptoms are due to mechanical causes then the heart might be saved. The surgeon is also concerned about whether rheumatic fever always affects the whole heart, as Sir James Mackenzie believed, or whether one part might bear the brunt: it would be useless to operate upon a valve if the myocardium was always destroyed as well. Information concerning these points is available in the writings of L. Gross. It is known that the brunt of the rheumatic attack can fall upon one part of the heart, leaving the remainder apparently untouched. Any patient who has had rheumatism may suffer from new attacks of the disease.

Paul Wood has stated that rheumatic carditis accounts for 25 per cent of all heart disease in hospital, and in half of these patients mitral stenosis was the chief lesion. It follows that if surgical treatment offers a reasonable chance of alleviating valvular sclerosis there is much work to be done. Indeed it has been calculated that there were about a quarter of a million patients of both sexes, between the ages of 18 and 44, in Great Britain. Practically all of these had one or more valve lesions; the mitral was said to be involved in 85 per cent, the aortic in 44 per cent, the tricuspid in 15 per cent and the pulmonary in 2 per cent. By no means all of these patients need surgical treatment, and many have passed beyond the possibility of relief. Rheumatic fever is becoming a less common disease than it was in the immediate past.

THE MITRAL VALVE

The surgery of the mitral valve comprehends the greatest part of cardiac operations today, and the problems which have been presented are being investigated in many countries. The term "mitral stenosis" has been used loosely in the past; it is now generally taken to mean any pathological deformity of the mitral valve in which there is an element of obstruction.

Surgical Anatomy. The mitral valve consists of a number of parts extending over a vertical distance of 6 cm. Each of these is essential to the normal function of the valve, and modern surgery demands a knowledge of the anatomy in health and disease. Here then are the anatomical facts about the mitral valve which concern the surgeon.

THE MITRAL RING is a fibro-muscular ring from which two cusps are slung. It is situated in the floor of the left atrium, and morphologically is a part of the primitive skeleton of the heart. The mitral ring contains blood vessels, but the leaflets of the valve are normally avascular. Blood vessels, in profusion, grow into the leaflets of the valve during episodes of rheumatic disease, and through them the noxious substances, which destroy the valve are carried to the membraneous parts of the leaflets. Throughout life

the cusps are constantly renewed from the ring in the same way as the nail grows from the nailbed. The normal aperture of the mitral ring is 6 sq. cm.

THE CUSPS. There are two flaps or cusps which hang from the mitral ring down into the ventricle. The *antero-medial* cusp is the larger and the more important: it is closely related to the aortic valve which is situated immediately in front of it and above it. The *postero-lateral* (mural) cusp is small and less mobile: it lies close to the muscular wall of

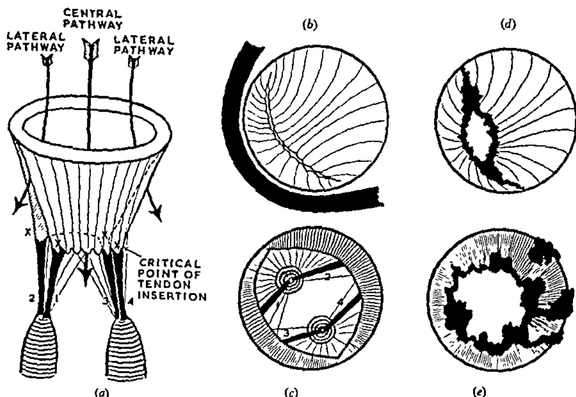


FIG 313. (*a*) Diagram depicting the parts of a normal mitral valve. (*b*) Mitral valve seen from above. Valve closed. The anteromedial cusp is larger than the postero-lateral mural cusp. (*c*) Mitral valve seen from above Valve open showing papillary muscles, chordæ tendinæ, and the points of critical tendon insertion (i e. 1, 2, 3, 4) (*d*) Mitral stenosis. Mainly an adhesive lesion. (*e*) Mitral incompetence A destructive lesion

the left heart, and functions principally as a cushion against which the antero-medial cusp is applied in systole. The two cusps are separate, not only at the lower margins of the valve but at the sides as far up as the mitral ring. The gaps between the vertical parts of the cusps are called the *lateral pathways* and the principal outlet through the valve is called the *central pathway*.

THE CHORDÆ TENDINÆ AND PAPILLARY MUSCLES resemble the cords of a parachute: they are tendons joining the cusps to the papillary muscles, which are two in number and part of the musculature of the left ventricle. When the ventricle contracts the apex of the heart is brought closer to the mitral ring above; the chordæ tendinæ are relaxed and this allows the anteromedial cusp to balloon against the mural cusp and so the valve closes. The function of the chordæ tendinæ is to prevent the cusps turning inside out after they have approximated.

Each papillary muscle is shaped like a small cone the size of an almond. From the top of this cone a number of tendons pass up to both cusps: these cords vary in strength

according to the place they are inserted into the cusps. The strongest and largest chordæ are about ¾ in. long and pass to the "critical area of tendon insertion," that is, to the point where the vertical part of the cusps turns at right angles into the horizontal lower edges. The less important chordæ pass to the lower edges and to the vertical parts on either side of the critical areas.

The effect of this arrangement of chordæ tendinæ is apparent if the normal valve be viewed from above. When the valve is closed the antero-medial cusp is apposed to the postero-lateral cusp and only a chink remains between the two: this chink, consisting of the central and the lateral pathways stretches straight across the mitral ring from 11 o'clock to 5 o'clock. When the valve opens the fairway is not round like the mitral ring because the chordæ tendinæ partially obstruct the lateral pathways leaving the central pathway patent, and rather square in shape. It follows that the lower outlet of the valve through the cusps is smaller than the inlet through the mitral ring. It also follows that any pathological process which causes deformities of the chordæ tendinæ and papillary muscles will destroy the function of the valve as surely as if it were in the cusps or the mitral ring.

Rheumatic disease may destroy the whole valve mechanism or any part of it: it may result in deformities which are incurable except perhaps by the provision of an entirely new mechanism, or in defects which can be repaired by simple procedures.

It is wrong to regard the mitral valve as a diaphragm which becomes contracted and which can be simply remedied by enlarging the obstructed hole. To act on this assumption is to convert mitral obstruction into acute mitral incompetence.

Surgical Pathology of Mitral Obstruction. In the acute phase of rheumatic carditis the cusps of the mitral valve become œdematous, and the lower edges, which are subjected to most of the trauma, are principally involved at first. The murmurs at this time are suggestive of incompetence due, probably, to the fact that the œdematous flaps do not come accurately into total apposition. Along the free margins of each cusp small thrombi develop and are particularly numerous along the edges of the pathways and at the critical areas of tendon insertion. As the œdema subsides the valve may be restored to normal, or sclerosis may develop In the latter event fibrous tissue obliterates the lateral pathways forming commissures on each side and a rubbery ring round the central pathway, i.e. the valve outlet. The destruction may include a deposition of calcium salts around the rim of the outlet and ultimately of large plaques of calcification throughout the cusps, the mitral ring, and the adjacent floor of the atrium. In other cases the sclerosis spreads downwards from the bottom of the cusps into the chordæ tendinæ and the papillary muscles which become transformed into blocks of scar tissue holding and fixing the flaps of the valve. Sometimes the whole valve from top to bottom is destroyed. There are differences of theory as to the details of how and when the commissures of the valve fuse together. Brock has stated: "*Fusion of the valve cusps . . . occurs fundamentally at the two opposing critical areas of tendon insertion.*" From these lower corners of the valve cusps, which stick together first, the adhesion spreads upwards along the lateral margins of the cusps to the mitral ring. Later on the adhesion may further reduce the central pathway or it may obliterate, scar, or fuse the chordæ tendinæ. Experience at operations has shown that in mitral obstruction the lateral pathways are always obliterated, and the areas of most dense adhesion are the critical points of tendon insertion. It is the original central pathway which remains patent; and thus *in mitral obstruction the size of the*

outlet is about the same in most cases, i.e. 1 cm. by 0·5 cm.; that is the space left unfused between the points of critical tendon insertion. The outlet may be smaller than this if the margins of the remaining pathway have been involved as well as the commissures.

According to this view of the development of mitral obstruction a patient who suffers from acute rheumatic carditis sustains certain pathological changes in the mitral valve at the time of the acute illness. These changes result in fusion of the commissures in some cases and this, which can occur early, has the effect of reducing the lumen of the valve to that of the central pathway. If the myocardium has not been importantly damaged by the acute carditis, and if the principal lesion has been in the mitral valve, then the heart may continue to do its work efficiently, in spite of the obstruction, and the patient may die of old age. If, on the other hand, the acute carditis has seriously injured the efficiency of the myocardium, the heart will fail straight away. But if, as usually happens, the patient recovers from the acute carditis, with only minor residual myocardial damage, an interval follows, during which some mitral obstruction is actually present, but during which the heart does its work without undue strain. In such a patient there may come a time, with advancing years and narrowing coronary arteries, or the sudden imposition of some extra strain such as respiratory infection, when the myocardium fails.

If this idea be true then the difference between a patient who has mild symptoms and another who is severely afflicted is not primarily connected with the size of the mitral outlet. It is a measure of the fact that in the severe case the myocardium can no longer compensate for the valve obstruction. It does not mean that the outlet is decreasing day by day.

The chief argument against this view is that it is difficult to understand how a patient whose valve is already obstructed could indulge in exercise—as many can—which would necessitate a raised cardiac output. Blood only flows from the atrium to the ventricle when there is a pressure gradient between the two chambers. If the outlet is constricted early in the disease to about 1·5 cm. × 0·5 cm. the pressure which must build up in the atrium to force the blood through would cause hypertrophy of the right ventricle and there is no evidence to support this in mild cases.

The most generally accepted view is that rheumatic carditis initiates a process which causes the commissures to be gradually obliterated over a period of years and that fusion occurs from the mitral ring downwards. The pathway through the valve is slowly and relentlessly shut down, and when a critical degree of obstruction has been achieved the signs and symptoms of mitral stenosis develop. It is presumed that after relief of the obstruction the theumatic sclerosis may continue as before, and the lumen could close up again.

The controversy described above has not been settled because patients with mitral obstruction do not die until the heart fails, but it matters to the surgeon for these reasons. If he believes that he is dealing with a longstanding mechanical obstruction, he will expect to get good results if the obstruction can be relieved without producing incompetence and if the myocardium is sound If he believes the second hypothesis his operation is not *likely to succeed for more than a short time, because he has operated upon tissues which* are still involved in a progressive pathological state.

Surgical Pathology of Mitral Incompetence. Regurgitation means blood from the ventricle is pumped back into the left atrium during systole. If one regards the pathological mitral valve as a diaphragm which has a hole in it, and rheumatic fever as a disease

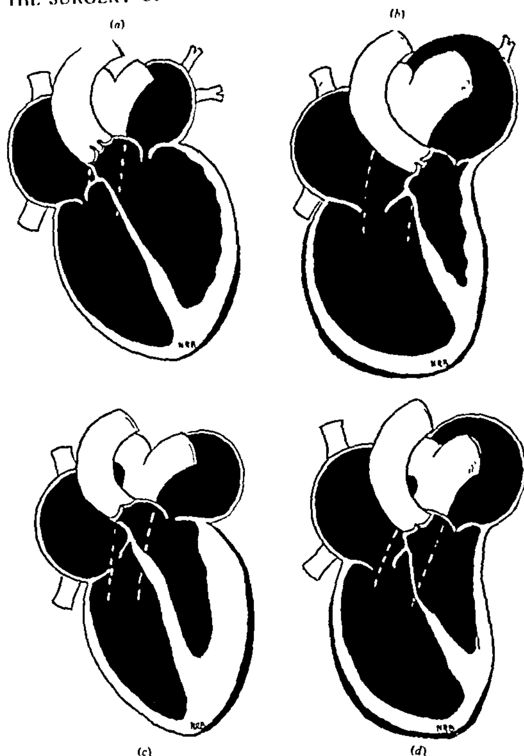


FIG. 314. Diagrammatic representation of the types of chamber involvement with various types of valvular stenosis (a) *Normal heart.* (b) The heart in isolated *mitral stenosis* Note the dilated left atrium and right ventricle The left ventricle is small; this chamber is protected, by the inlet stenosis, against strain (c) The heart in isolated *aortic stenosis*. The severe stenosis at the outlet of the left ventricle causes hypertrophy of the musculature of this chamber The left atrium and the right ventricle remain normal until left ventricular failure is immanent. (d) The heart in combined *mitral and aortic stenosis*. Note the similarity to the condition found in pure mitral stenosis The right ventricle and left atrium become dilated and overworked; but the left ventricle remains small and relatively healthy. It is protected against the effects of stenosis at its outlet by the severe inlet stenosis The small amount of blood entering the left ventricle is readily expelled through the narrowed aortic valve.

(These diagrams have been copied from a paper on aortic stenosis by Bailey of Philadelphia)

which diminishes the size of that hole: then it is natural to visualize the orifice contracting down, and to think of mitral incompetence as a step on the way to mitral stenosis: a less severe type of mitral obstruction. *This is untrue.* There are a number of causes of mitral incompetence: but most are the late result of rheumatic disease. Incompetence generally occurs when the whole valve, from the mitral ring to the papillary muscles, has been destroyed. In mitral obstruction the abnormality generally affects the free edges of the valve cusps which fuse together; the membraneous parts remaining supple and mobile at first. In mitral incompetence the brunt of the disease falls upon the tissue of both cusps and the mitral ring. The result is that, when scarring is followed by contraction and calcification, the edges of the cusps around the central pathway retract towards the mitral ring and enlarge the pathway. To make matters worse the chordæ tendinæ are shortened so that the valve is reduced to a rigid diaphragm in the middle of which is a large hole with craggy margins. Thus mitral incompetence is a more severe deformity than mitral obstruction.

From what has been described above it follows that:

(1) When adhesion between the edges of the cusps is the major lesion "pure" mitral stenosis results.

(2) When contraction and destruction are the major lesions "pure" mitral incompetence results.

(3) When adhesion and contraction combine mixed valve abnormalities result.

Obstruction and Incompetence frequently occur together in the mitral valve. If obstruction is severe there is no important regurgitation, and if regurgitation is gross there is no obstruction: but in many cases the central pathway is not only narrowed but its margins are held open. How much regurgitation occurs as a result of this, depends not only upon the size of the patent fairway but upon the direction in which the orifice points in the ventricle. If it so happens that the deformed valve is held by scarred chordæ so that the opening faces the stream of blood which should be directed up the aorta, an important part of that stream will pass back into the atrium. Brock has stated that regurgitation occurs in three forms:

(i) A tiny jet of blood which flows back through a tight and fixed mitral obstruction and is of no significance.

(ii) A moderate reflux through a valve which is not grossly obstructed, the central pathway being perhaps 3 cm. in transverse diameter. This is the usual form.

(iii) A powerful reflux which deflects most of the blood back into the atrium. The mitral aperture in such a case may be 4 cm. × 2 cm., and the atrium aneurysmal.

The Effects of Mitral Disease. If the disease which destroys the mitral valve results in pure obstruction, the pathological and clinical effects are confined to those parts of the circulation proximal to the valve. If, on the other hand, regurgitation is the principal defect the left ventricle and the systemic circulation are also affected. In theory mitral obstruction and mitral regurgitation are so different that accurate diagnosis, as between the two, should be possible. In practice they are not always clearly separated.

In all types of mitral disease the left atrium enlarges and may become aneurysmal. The direction of the enlargement is backwards and towards the right, so that, in these patients it is possible to enter the left atrium from the right chest if circumstances dictate such an approach. This enlargement is not solely due to progressive increases in the pressure within the chamber: it is often the direct result of rheumatic carditis which has

particularly affected the muscle. The dilatation is greatest in mitral incompetence, and sometimes the movements of the atrium observed on screening become paradoxical. The course of the disease is often relatively benign in patients who have massive atria. As the atrium and the auricular appendage dilate, clot is apt to form in its relatively stagnant byeways and to become adherent to the walls. This complication is most usual when auricular fibrillation has occurred and, by engendering embolism, it adds importantly to the risks of the disease and of surgical treatment.

The pulmonary veins become distended and tense.

The changes which occur in the lungs in patients suffering from mitral stenosis are very important to the surgeon. If the operation he does to overcome the obstruction in the valve is successful, the clinical result may yet be a failure if the pulmonary hypertension does not subside. Numerous investigations have shown that, as time passes, the following changes occur in the lungs. The alveolar walls become œdematous; the basement membranes of the alveoli become thick and lined in places with cuboidal cells; the lumen of the pulmonary arteries is reduced by muscular hypertrophy of the media, and there is arteriolar necrosis in the arterioles. In addition all patients suffering from mitral stenosis show some degree of hæmosiderosis in the lungs; the cause of this is uncertain but it may be associated with the numerous small interstitial hæmorrhages which occur, or it may be a result of pulmonary hypertension. The radiographs of these patients show not only pulmonary plethora, but, upon occasion tiny spicules of bone formation in the lungs and horizontal linear shadows in the lower parts of the chest which are due to œdema in the interlobular septa and dilatation of the lymphatics upon the surface. When the chest has been opened at operation these lungs look purplish in colour, they are often difficult to deflate, and they may feel œdematous.

As the pressure in the pulmonary capillaries rises the patient becomes liable to attacks of pulmonary œdema. Theoretically these occur whenever the pulmonary capillary pressure exceeds that of the colloid osmotic pressure of the blood, and are particularly liable to start whenever a sudden increase in the output of the right ventricle occurs—as for example in tachycardia. During tachycardia the diastolic drainage time from the left atrium is reduced and the right ventricular output is possibly increased.

Hæmoptysis, dyspnœa, and pain caused by pulmonary infarction, are sometimes also directly due to pulmonary hypertension.

The pressure in the pulmonary arteries is always high and the vessels themselves may be greatly enlarged and white in colour. The right ventricle hypertrophies and eventually dilates, and the tricuspid valve may become incompetent. Back beyond this the pressure in the venæ cavæ rises, the liver enlarges and becomes tender. Ascites and œdema and pleural effusions occur and add to the embarrassment of respiration.

It was formerly believed that all the blood pumped out of the right ventricle must of necessity reach the left, but it is probable that a safety valve or buffer exists in the bronchial veins. These veins are particularly developed in patients suffering from mitral obstruction, and, when the pressure in the left atrium rises, a proportion of the blood could be diverted back to the azygos system.

The Natural History of Mitral Disease. Several large series of patients have been followed for 20 years or more. Recently Olesen has studied 351 cases of mitral disease of whom 271 suffered from pure mitral stenosis. He makes the following observations. The ages at which the disease of the valve was first declared varied from 14–73 years. There

was a previous history of rheumatic disease in 58 per cent. The first cardiac symptoms developed, on the average, 14 years after the attack of rheumatic fever, that is in the early thirties. The length of life of the patients after the heart disease was manifest varied from a few months to many years but averaged about 18 years. The average age of death was 47.

Some patients suffer from repeated attacks of rheumatic fever. The development of fibrillation is often the turning point in their careers, and marks the moment when deterioration begins. By contrast some suffer from severe mitral stenosis and die of old age. Isolated attacks of hæmoptysis are not necessarily of bad prognosis; but once emboli occur they are apt to be repeated and to influence the prognosis adversely.

The unknown factor in this story is the presence or absence of active rheumatism, and there is no easy way of deciding this point clinically when definite signs are absent.

MITRAL STENOSIS

Diagnosis. The principal symptom is increasing dyspnœa on exertion. Dyspnœa is not due to oxygen lack but to pulmonary congestion. It occurs not only when the patient takes exercise, but in paroxysmal attacks for which emotional upsets are often the initiating cause; it is influenced by changes in posture, and severely disabled patients can only sleep in the sitting position. Hæmoptysis, which can be profuse, and may be the first symptom, is also due to pulmonary hypertension or emboli or thrombosis. It occurs in about 50 per cent of cases. Most patients are subject to attacks of acute bronchitis which accelerate the progress of the disease. Episodes of acute pulmonary œdema are common, and occur most often in the early phase, before the right ventricle has begun to fail. Congestive heart failure complicates one in five cases. Pain of the anginal type sometimes occurs but pain due to pulmonary infarction is common, and systemic embolism is found in 10-15 per cent of cases.

In a typical case there is a malar cyanotic flush and rather cold extremities. The pulse is small but strong: it is generally regular in the early stages, but auricular fibrillation *occurs in 50 per cent of cases at some period in the illness.*

There is a systolic lift over the right ventricle, but the left ventricle is normal: the left atrium, the pulmonary artery, and the right heart enlarge. The first heart sound is accentuated. An "opening snap" may be heard in diastole. This sound which occurs at the moment the pressure in the atrium exceeds that in the ventricle, is an important sign; it may be heard in most cases of mitral obstruction, and when present, it is unlikely that incompetence is the dominant abnormality at the mitral valve. There is a diastolic murmur in the mitral area which may be accompanied by a thrill on palpation and which may be accentuated in the presystolic phase. There is a loud, pulmonary, split, second heart sound at the base. The lungs show the physical signs of plethora. Tight obstruction can exist when no opening snap and no diastolic murmur can be heard.

The appearances on screening are suggestive. The heart may be enlarged. The aorta is normal, but the pulmonary artery is large: the left atrium is large (the size of the atrium is best demonstrated by a barium swallow), the right ventricle is enlarged and the lungs are congested. Calcification can often be seen in the mitral valve itself or in the floor of the left atrium. The electrocardiogram shows right-sided preponderance and larger than normal or bifid P waves.

It has been established that the pressure in the pulmonary artery is raised in both

obstruction and incompetence; it bears but little direct relationship to the size of the orifice in the valve. It is also known that by passing the tip of a cardiac catheter onwards through the pulmonary artery, until it impacts in one of the smaller arterioles, pressure tracings can be secured which are identical with those in the left atrium of the patient.

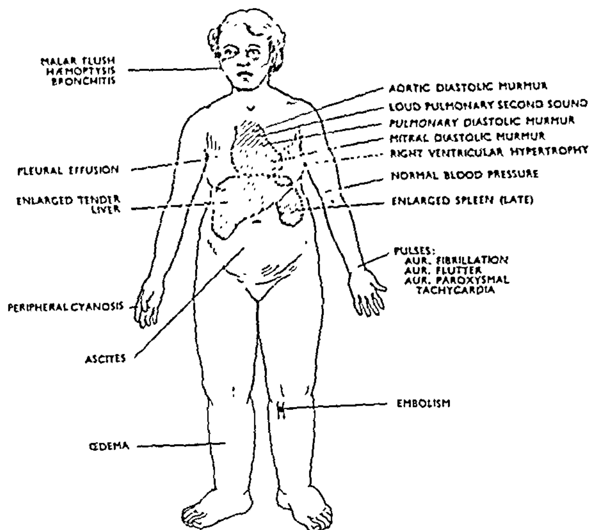


FIG. 315. Mitral stenosis.

The readings are called pulmonary capillary pressure tracings. They often fail as a guide to exact diagnosis.

In attempting to assess what weight to place upon the signs of mitral obstruction and mitral incompetence remember that there are typical signs of mitral obstruction, and of mitral incompetence. The signs of incompetence are apt to be masked by the signs of obstruction when both are present at the same time. The pulmonary capillary pressure tracing of mitral incompetence is probably typical of the condition, but when the left ventricle is failing the pulmonary capillary pressure is similar to that found in mitral obstruction.

When the diagnosis as between mitral stenosis and other obstructions in the left atrium is in doubt—i.e. myxoma of the left atrium on congenital stenosis of the pulmonary vein (cor triatrium)—the right side of the heart should be catheterized to

verify that the pressures in the pulmonary artery and in the pulmonary capillaries are high. And in addition the pressures in the left side, together with the pressure gradient across the mitral valve itself, should be measured. The latter can be done either by passing a long needle down a bronchoscope and inserting this needle straight through the wall of the main bronchus into the left atrium (Allison) or by passing the needle through the chest wall into the left atrium (Brock). In both cases the pressures must be recorded electrically.

Indications for Operation. There is no reason to operate upon a patient who has mitral obstruction without disability: such a one may reach mature age. The danger signal is the development of progressive limitation of exercise tolerance, embolism, nocturnal dyspnœa, or congestive failure. In general, patients under the age of 20, who may still have rheumatic fever, and those over 60 are unsuitable for surgical treatment.

The *ideal patient* for surgical treatment has the typical clinical signs of mitral obstruction, uncomplicated by other valvular lesions or by myocardial insufficiency. The signs *should suggest that the cusps of the valve are mobile*; that is an *"opening snap" should be* present. The left ventricle should be of normal size, the pulmonary arteries large, and there should be right ventricular preponderance on electrocardiography. The presence of some calcification in or near the valve, as seen on screening, does not give information as to whether operation will succeed or not; but massive calcification is of poor augury. There should be no evidence of active rheumatism.

Auricular fibrillation and a history of previous embolic episodes are points in favour of early surgical intervention. In both, the risks of surgery are increased but so are the risks of conservative treatment. When mitral obstruction is complicated by disease in the aortic valve the justification for operation is that the mitral lesion is considered to be responsible for the most threatening symptoms. Mitral obstruction, by itself, should no longer be regarded as a reason for terminating a pregnancy or sterilizing a young woman.

The Risk of Operation. The danger of operating depends upon the severity of the cases accepted, but the mortality in hospital should not exceed 6 per cent; it will be less if severe cases are refused.

The absolute contraindications to operation diminish as experience increases. These patients have obstruction, and many apparently hopeless cases can be relieved if the obstruction can be overcome. At the moment the following cases are bad surgical risks: those who have intractable cardiac failure and a large heart, particularly young subjects; those in whom the weight of the disease has fallen on the aortic valve but who have mitral stenosis as well; those who have signs of rheumatic fever; and those who have severe pulmonary hypertension. A valve which, at screening, is heavily calcified may split easily at operation, but a lot of calcification suggests a rigid and incompetent valve. It is unfortunate that there is no clinical method of assessing the degree of myocardial disease in these patients; but it is apparent that if, in fact, the brunt of the damage has fallen upon the myocardium then operation upon the mitral valve will not always succeed. On the other hand follow up results have shown that a successful valvotomy generally produces a good and lasting result. Experience has shown that death on the table is generally due to technical mistakes, such as hæmorrhage, anoxia leading to ventricular fibrillation, acute mitral incompetence due to avulsion of chordæ tendinæ or faulty splitting of the valve.

Pre-operative Treatment. Patients suffering from congestive failure at the time operation is contemplated, should be treated medically in the hope that this may be relieved. Full digitalization, a low salt diet, abdominal paracentesis, Southey's tubes to drain off the œdema, mercurial diuretics, rest, and moderate dehydration help a great deal. If pleural effusions are present they may need aspiration before operation: breathing exercises should be given. Sedatives may be used freely and with advantage.

Attacks of acute pulmonary œdema require special consideration In general, they are an urgent indication for operation for in no other way can they be cured. They occur particularly in patients who cannot increase their cardiac outputs. If, during preparation for operation, attacks of pulmonary œdema occur, they must be energetically treated as follows. The patient should be sat up, and the legs put down. Venous tourniquets should be applied to the upper parts of the legs and to the arms to diminish venous return; venesection may be performed (300 ml.) in urgent circumstances: aminophylline (gram 0.48) intravenously is valuable; morphine (grain ¼) and oxygen are essential. No patient should be allowed to drown in his own bronchial secretions; these should be sucked out as necessary by intratracheal intubation or bronchoscopy. It sometimes happens that a patient goes into an attack of pulmonary œdema just before anæsthesia is induced. The surgeon and anæsthetist must then decide whether to proceed or to delay operation. If the surgeon elects to operate and fails to overcome the obstruction the patient will probably die; on the other hand there is no permanent cure for the condition except by operation. If the patient has already had several attacks it is reasonable to hope that he may survive another, and operation is put off. To avoid a repetition of the œdema at the next attempt the patient may be more heavily sedated before coming up to the theatre.

The Principles of Surgical Treatment. To date two different methods, indirect and direct, have been advocated to treat mitral stenosis. The indirect way is to prevent some of the blood pumped from the right heart from reaching the left atrium, and so to diminish pulmonary congestion. Sweet of Boston tried to do this by *anastomosing a branch of the right inferior pulmonary vein to the azygos vein.* This method has not been generally used, not only because of the technical difficulties; but because the anastomosis, being between two veins, tended to clot. A modification of this idea was *ligation of the inferior vena cava.* The ligature was placed immediately below the renal veins. The operation, which is simple, is done through an incision similar to that used for lumbar sympathectomy and keeping behind the peritoneum. It affords some patients, and particularly those with right sided heart failure, temporary alleviation of symptoms: the œdema of the legs and the ascites are improved.

The present technique involves operating directly upon the *mitral valve.* The operation is done blindly with "the eye on the end of the finger," and as it stands today, it is but a milestone in the progress of surgery. Nevertheless the technique, which was first devised by Souttar, has already proved beyond doubt that intracardiac surgery is possible and desirable, and that rheumatic valvular disease in particular can often be alleviated.

The ultimate aim is to devise methods of operating under direct vision, and to replace valves, which have been destroyed, by grafting in new cusps. The operation of mitral valvotomy is a good stopgap.

The most important point about valvotomy, as it is performed today, is that success depends not merely upon enlarging the fairway through the obstructed valve, but upon

doing this without producing mitral regurgitation. Thus the object is more complicated than that of punch prostatectomy with which it has been compared.

The earliest attempts of Cutler and Beck, which were carried out through the left ventricle, succeeded in overcoming the obstruction by punching a piece out of the antero-medial cusp; this led to acute incompetence, and the patients died. The present operation rests upon the idea that if the two commissures can be divided, from the central pathway to the mitral ring, the obstruction will be overcome and the cusps of the valve will be freed to function once again. Any patient in whom both these objectives are not achieved will get a poor clinical result.

Technique of Mitral Valvotomy or Commissurotomy. The operation is done under general anæsthesia and a blood transfusion is set up in case of hæmorrhage during the intracardiac manipulations. The chest is opened through a long, left, lateral incision through the fourth interspace and reaching from the edge of the erector spinæ at the back to the costal cartilage in front. The lung is generally of a purplish hue, it feels doughy and can only be deflated slowly by retraction. Pleural adhesions may require division. Having exposed the pericardium, this sac is opened by an incision passing downwards from the top of the pulmonary artery to the bottom of the left atrium. It is unnecessary and undesirable to carry the opening too far over the ventricle. At this point it can generally be ascertained whether the heart is irritable or not, by gentle pressure at any point upon the surface: if such pressure, or the application of a small cold swab, causes ectopic beats the surgeon and the anæsthetist must decide whether procaine should be given intravenously or whether a weak solution of some cocaine derivative should be applied to the surface of the heart. On the whole it is unnecessary to use these drugs and it is more important to avoid arrhythmias by doing the operation skilfully and expeditiously than it is to endeavour to damp them down with drugs. On opening the pericardium the left atrium and the auricular appendage are exposed. In most cases the pericardium is normal but sometimes it is obliterated by adhesions, some of which must be separated to get access to the appendage. The pulmonary artery is enlarged and the pulmonary veins are distended.

Experience has shown that, as a result of operations upon the mitral valve, there is a risk of *embolism*, due to fragments of clot or calcified particles being dislodged from the atrium or from the margins of the valve during the intracardiac manipulations. Tiny fragments cause damage if they lodge in the cerebral vessels, and large masses of thrombus, which cannot get to the brain because of their size, may obstruct peripheral vessels. It is not possible to predict, in any individual case, whether embolism is likely to occur or not. The masses of organized thrombus which are sometimes found in the left atrium are generally firmly adherent to the walls of the chamber and seldom cause trouble; whereas the newly formed, friable, clots, which may not be seen or felt at operation, are dangerous. The surgeon is thus in somewhat of a quandary as to how to deal with this problem of threatened cerebral embolism he can do little at valvotomy to avoid aortic or peripheral limb emboli. Two different techniques to avoid cerebral embolism are available; the majority of surgeons deliberately amputate the tip of the auricular appendage before applying a clamp to the base and, by letting the blood flow out freely, they hope that any loose pieces of clot will be washed out of the atrium. Others prefer to expose the three large branches which come off the aortic arch in the mediastinum (in about 8 per cent of cases the left vertebral artery comes off the aortic arch as well as the other vessels) and

to clamp them all off completely whenever any active manipulations are undertaken inside the atrium or at the valve. The cerebral vessels can be safely occluded at repeated intervals but not for longer than a minute at a stretch. Using the latter technique the writer has overcome the problem of cerebral embolism in a large consecutive series of cases. The possibility of an embolus having lodged at the aortic bifurcation or in one of the leg arteries is met in this way: before operation all the pulses in both legs are palpated; if, at the conclusion of the operation the blood supply to one or both legs has changed the desirability of doing an immediate embolectomy must be weighed. These points about embolism are discussed at this time, because the surgeon who treats these patients must apply his own chosen plan of campaign at this point in the operation.

Having safeguarded the patient against the risks of embolism the auricular clamp is tightened and the inside of the appendage distal to the clamp is examined. In most cases strands of muscle, columnæ carneæ, will be found passing from one side of the appendage to the other and these must be divided before attempting to introduce the finger. If the appendage contains organized clot this must be separated from the wall and removed. Purse-string sutures around the base of the appendage are unnecessary, unless the appendage is so small that the only way of getting into the atrium is by carrying the incision in the appendage through the wall of the atrium as well. If a purse-string stitch is necessary it is convenient to control it by threading the ends through a special flexible tourniquet. An auricular appendage of average size will admit an index finger and the narrow neck of the appendage fits the finger snugly and prevents bleeding. Some few cases occur in which access through the appendage is impossible: in such the little finger can sometimes be introduced through one of the pulmonary veins, but this technique is more dangerous than cutting into the wall of the atrium from the base of the appendage and using a purse-string stitch to control the incision. The index finger is passed into the atrium by inserting the tip into the appendage and removing the clamp. It is as well not to lubricate the finger, with liquid paraffin for example, because tiny drops of oil might cause cerebral embolism. Some surgeons prefer to remove the glove from the operating finger because it makes palpation inside the heart more accurate. Once the finger has been introduced into the atrium the pathological anatomy of the mitral valve is assessed by palpation, taking care not to obstruct what remains of the pathway between the atrium and the ventricle. [illegible]

the central pathwa[illegible]

way once again reaches right out to the mitral ring on each side. In other cases the margins of the central pathway feel like a ring of tough indiarubber and, in these some form of commissurotomy knife must be introduced on the flexor aspect of the surgeon's finger. With this knife the two critical points of fusion are cut through and, when this has been done, it is generally easy to divide the commissures out to the mitral ring with the finger alone. Occasionally the valve is so destroyed by scarring of the cusps and fibrosis of the chordæ tendineæ that no operative intervention is possible: to struggle against the impossible in these circumstances invites such accidents as perforating the ventricle or avulsion of a papillary muscle. It is unusual for calcification to be the principal cause of inoperability: in many of the most favourable cases there are fine spicules of calcification along the rim of the central pathway, and sometimes the commissures split easily in spite of large bosses in the cusps. Bailey stated that the commissures should be split until the apical diastolic thrill disappears on direct palpation of the

heart. Commissurotomy should not result in a regurgitant jet palpable during ventricular systole; if it does the late result of operation is not likely to be good. After commissurotomy the finger is withdrawn and the clamp reapplied to the base of the auricular appendage. The incision in the appendage is closed, using as few stitches as possible, and placing them in such a way that uneven crevices, in which thrombi could form, are not left between the stitches. If the incision in the pericardium has not been too big it is not sewn up because it is wise to drain the postoperative pericardial effusion into the pleural cavity. Do not put antibiotics into the pericardium and sew it up tightly. Temporary pleural drainage should be provided and the surgeon may or may not leave antibiotics in the pleural cavity. If the pressure in the left atrium has been taken before and after commissurotomy there should be a significant drop.

Alternative Procedures at Operation for Mitral Stenosis

The Incision. Most surgeons use a long, left postero-lateral incision to approach the mitral valve. This gives good access to all parts of the left heart and to the superior mediastinum. Some prefer a left antero-lateral approach, reflecting the breast and pectoral muscles upwards. Bailey has recommended a *right* thoracotomy; in these cases the left atrium enlarges backwards and to the right. By dissecting the right pulmonary veins backwards and defining the back of the interatrial septum a good exposure of the left atrium may be had. Using this approach the tricuspid valve can be palpated (by opening the right auricular appendage) as well as the mitral valve.

Approach to the Mitral Valve

Although the conventional approach is by way of the left auricular appendage, some surgeons, and particularly Logan, advocate a left ventricular route in most cases. The valve aperture is palpated in the usual way through the left auricular appendage and, if the commissures cannot be split digitally, an incision is made about 2 inches below the mitral ring into the chamber of the left ventricle. Through this hole a special expanding dilator is passed into the ventricle and guided up through the stenosed orifice by the tip of the finger in the atrium above. An excellent commissurotomy is possible in difficult cases using this method. It has the additional advantage that the instrument nearly always deals effectively with the postero-medial commissure which can be difficult from above. Dubost, of Paris, has advocated using an expanding dilator passed from the auricular appendage downwards. Whenever a mechanical dilator is used there are two theoretical risks; the expanding blades may get entangled in the chordæ tendinæ so that it is hard to get the instrument out of the heart, or the dilator may tear a cusp in the wrong place and disorganize the whole valve mechanism.

Patients Suffering from Aortic and Mitral Stenosis

When both valves are obstructed they can be dealt with effectively at the same operation. Before embarking on the double valvotomy it is wise to make sure that the aortic valve needs opening. Unlike the mitral valve it is difficult and dangerous to palpate the aortic cusps, and the answer lies in taking the pressures in the left ventricle and the aorta simultaneously with an electric manometer connected to needles inserted at the appropriate places. If there is a pressure gradient across the aortic valve of more than 20 mm. of Hg. an aortic commissurotomy should be done. The mitral valve should be done first.

The surgeon may elect to cut the mitral valve through the left auricular appendage and then open the ventricle to get at the aortic valve; but most prefer to do both valves through the same incision in the left ventricle. A special adaptable dilator (Tubbs) is necessary for this purpose, as the size of the aperture to be achieved is different in the two cases.

Convalescence. On return to the ward the patient will need sedatives and perhaps oxygen therapy for a few hours. A bloodstained effusion, amounting to less than a pint, will collect in the pleural drainage bottle during the first 24 hours, and after this pleural drainage can be discontinued. Most patients are convalescent within a few days and feel the benefit of the improvement in their circulations straight away. Dyspnœa is rapidly relieved and the heart does not "thump when it beats." The physical signs in the heart rarely return completely to normal. Digitalis, which will have been exhibited before operation, must be continued for 4–6 months afterwards; and the patients must be warned not to test the benefits they have received too early or too actively in convalescence. In particular attacks of bronchitis or catching a cold may cause a setback. Most patients who suffered anginal pain before operation will be completely relieved of this symptom.

The Complications of Operation

Pyrexia

Most patients develop a transient pyrexia after operation, and if this be associated with malaise, an increase in the venous pressure, and tachycardia, some anxiety may be felt. These symptoms were at first unexplained and were called "the commissurotomy syndrome"; they are almost certainly due to traumatic pericarditis, and resolve without treatment. They are most obvious when antibiotics have been left in the pericardium.

Transient jaundice is not uncommon and is of no significance: the liver may be tender for several days.

Pyrexia and joint pains suggest a recrudescence of rheumatic activity and should be treated by adequate doses of salicylates.

Abnormalities of Rhythm

If auricular fibrillation was present before operation, it is likely to continue afterwards but a few of these patients can be reverted to normal rhythm in the early post-operative period. In some cases, where the rhythm was normal before operation, fibrillation develops during the first week of convalescence. These should be treated with adequate doses of digitalis and quinidine, and if normal rhythm results quinidine and digitalis must be given for many months to avoid relapses.

Cerebral Embolism

The most serious complication is cerebral *embolism*. It generally occurs during the operation and unknown to the surgeon, but cases of delayed embolism occurring during convalescence have been reported.

The severity of cerebral embolism varies. Some patients never recover consciousness and die after one or two days; in others the lesions though serious at first are transient and minor. The surgeon can never be sure that cerebral embolism has occurred until several hours after the operation, but the condition may be suspected if recovery from anæsthesia is delayed and the pupils remain inactive. The difficulty lies in the fact that many of these patients have abnormal hepatic function, they detoxicate anæsthetic agents

slowly, and slow recovery from anæsthesia may be due to this cause. If the patient who has had an embolus does not get progressively worse and die within a few days, the chances of recovery are good. To quote but one example: a patient who was unconscious for a week, is now sufficiently recovered to lead a reasonably normal life. A patient who has been severely paralysed will be left with some residual weakness, but prognosis in the early stages is impossible. A possible treatment is to reduce cerebral œdema and improve the collateral circulation by blocking the stellate ganglion. During convalescence re-education is important. The real treatment of cerebral embolism is to avoid it at all costs.

Peripheral Embolism

At the end of every operation the pulse should be felt in the legs and in the arms. If a pulse, which was palpable before operation, can no longer be felt, and the limb is cold, it is probable that embolism has occurred and embolectomy should be carried out immediately.

Pulmonary Infarction

This is another complication which is common in its minor forms. Many of these patients have been bedridden for a long time and if the signs of thrombosis in the legs become apparent they should be given anticoagulants in the usual way. Pulmonary infarction may also be due to embolism from the right auricular appendage, the right atrium, or to thrombosis in a branch of the pulmonary artery. The latter is most likely to occur when atheroma of the pulmonary arteries is present. Pulmonary infarction is often found soon after the onset of auricular fibrillation, which suggests that a thrombus in the right side of the heart may be responsible. It can be a cause of chest pain, hæmoptysis, and sometimes, of pleural effusion. In some cases pulmonary embolism is repeated in spite of anticoagulants, and then vein ligation will be necessary to save life.

Electrolyte Imbalance and Suprarenal Failure

Harken has pointed out that some patients suffer from water retention after operation, and that this, combined with suprarenal exhaustion, may be sufficient to weight the scales against a successful result.

Results of Operations for Mitral Stenosis. Several series of patients operated upon more than 5 years previously have now been reported. The outstanding point in all these papers is that if the surgeon has succeeded in doing a really efficient commissurotomy the result is likely to be good and lasting. This observation supports the view that obstruction at the mitral valve, and not myocardial disease, is the dominant lesion in this disease.

The second feature which controls clinical success or failure is the degree and extent of pre-operative pulmonary hypertension. There is, as yet, no accurate knowledge as to how much, and under what circumstances, this type of circulatory obstruction can return to normal.

The size of the heart, auricular fibrillation, hæmoptysis, and limitation of exercise tolerance are not factors which militate against a good result. A heavily calcified valve is not likely to be made capable of normal function by operation.

The early results of mitral valvotomy are that about 60 per cent of patients are dramatically cured; in 30 per cent the subjective symptoms are much improved but some signs of obstruction remain; 10 per cent are failures.

Detailed follow up investigations have shown that as the years pass the numbers in each of these categories change so that some of the patients who appeared to have been cured slip into the class of those who have residual signs of obstruction. Some die and some again develop all the signs and symptoms of mitral stenosis. The relevant figures quoted by Brock (1955) are as follows: 2 per cent lost all the improvement in the first year; 6 per cent in the second year; 10 per cent in the third year and 15 per cent in the fourth year. On the other hand these were all early cases, and it may well be that since adequate to perfect commissurotomy is now achieved in a much higher percentage of cases, the late decline in the good results will be checked.

Recurrence of Mitral Stenosis

In about 50 per cent of all atrial appendages amputated at operation on the mitral valve, the characteristic lesion of rheumatic fever, the Aschoff node, is found, no matter how remote the original attack of rheumatic fever (if there has been one). This finding suggests that the active process is continuous though, unsuspected, in many cases; if this is so then in a proportion of cases the mitral valve obstruction will recur, even in the absence of attacks of rheumatic fever. And it is now certain that many patients whose signs and symptoms have recurred will be submitted to a second operation. Many such operations have already been done. The immediate surgical risks are higher than at the first procedure but good results can be achieved. In these cases the pleural and pericardial cavities are generally obliterated by adhesions and landmarks may be difficult to find. The fact that the auricular appendage has already been amputated generally means that the atrium can only be opened by using a purse-string suture and a tourniquet. In some cases the surgeon may elect to operate from the right side of the chest and to expose the large left atrium by reflecting the right pulmonary veins backwards. This approach affords good access not only to the mitral valve, but to the tricuspid valve as well.

MITRAL INCOMPETENCE

It has already been described how mitral incompetence can coexist with mitral obstruction, and that both defects may sometimes be improved by commissurotomy. The present remarks refer to those patients in whom incompetence is the principal defect, and for whom the operations described above are of no avail.

The causes of mitral incompetence are:

(i) The patient may have a defect in one of the mitral cusps. In this connection, as explained above, the anteromedial cusp is more vital than the postero-lateral.

(ii) The valve cusps may be so shrunken by disease that the edges cannot meet.

(iii) The musculo-tendinous mechanism may be ruptured by trauma or bacterial infection or operation.

(iv) The musculo-tendinous mechanism may be so scarred and contracted that the cusps *cannot* shut.

(v) The mitral ring may be congenitally or pathologically large.

The Diagnosis as between obstruction and incompetence was considered straightforward until surgeons proved that the cardinal signs of both these conditions might not

be pathognomonic. Nevertheless there are certain signs which are important, but the diagnosis should never be made on any one sign; it rests upon a combination. The history is usually longer than that of mitral obstruction. Incompetence occurs 5 times more commonly in men than women.

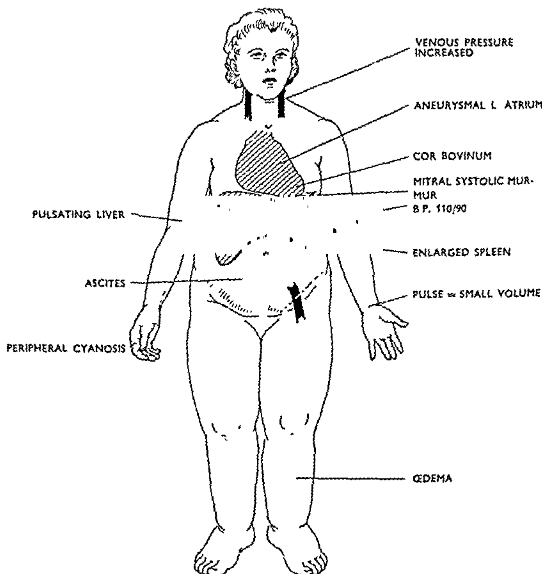


FIG. 316. Mitral incompetence

In mitral incompetence it is typically the left ventricle more than the right which is enlarged, and this is decided by palpation and screening: but principally by electrocardiography. The first heart sound is soft at the apex. There is an apical systolic murmur which extends up to the second sound and is conducted into the axilla. There is rarely an "opening snap." The most important radiological signs are said to be systolic, or paradoxical, expansion of the left atrium, and great enlargement of the left atrium.

The surgeon should be suspicious of incompetence in a patient, said to be suffering from mitral stenosis, who has a large left ventricle. The common causes of a large left ventricle are hypertension, aortic valvular disease, mitral incompetence, and myocardial disease: mitral obstruction is not one of them. In a difficult case the diagnosis can only

be made at operation, for it is then that the surgeon palpates the open fairway and feels the strong spate of the regurgitant stream. A surgeon who operates for mitral obstruction may find mitral incompetence. Some cases may eventually be helped by operation.

Treatment. At the present time the surgical treatment of mitral incompetence is in the experimental phase although many different operations have been tried.

In patients suffering from a large mitral ring, in whom there is no obvious disease of the valve, Bailey and others have recently tried to reduce the size of the ring by inserting lasso stitches of pericardium or of fascia. This was not a new idea. Cushing and Branch in 1908 narrowed the tricuspid and mitral valves of dogs by inserting a silk ligature, and in 1909 Berheim produced mitral stenosis in this way. Thrombosis around the suture was the chief cause of death in these animals. In 1930 Wilson produced mitral stenosis in dogs by inserted flaps of pericardium across the mitral pathway: he found that these flaps were relatively easy to introduce and that they did not cause clots to form. By contrast, fascia lata, introduced in the same way, did produce clots and the fascia disintegrated within a few weeks. Murray and others in 1938, and again in 1950, replaced the postero-lateral mitral cusps in dogs, and later they did the same thing in patients, and found that it was possible to substitute a graft formed of a superficial vein turned inside out over a tendon. From these experiments it appeared that flaps of pericardium could be introduced across the left ventricle without undue risk to the heart.

In mitral incompetence the desirable operation is to introduce a new valve accurately and under direct vision. This is not possible at the moment, but two makeshift operations have been tried, with some success.

Logan, in Edinburgh, and several others have used pericardial flaps with some clinical advantage. The heart is exposed, as described above, and the diagnosis is made, or confirmed, by the finger in the atrium. A large flap is cut from the anterior pericardium and rolled into a tube pedicle. The finger is once again passed through the open mitral pathway, and the free end of the pericardial tube is drawn through the chamber of the left ventricle and out at the back. The tube pedicle is directed through the chordæ tendineæ by the finger in the heart so that it lies just below the patent mitral pathway, and it is drawn just so taut that it can flap down in diastole and up in systole. In this way a flap valve is provided. How long such a flap lasts is a matter for speculation. Logan has modified his technique by passing the pedicle of pericardium across below the valve as a sheet of tissue which is anchored in several places.

In these patients the postero-lateral cusp is almost totally deficient and the remnant of the important antero-medial cusp has no cushion to contact. If, at operation, the finger is passed through such a mitral valve one can sometimes feel the aortic cusp touching the front of the finger during systole, and when this happens the peripheral circulation of the patient improves. It follows that such a patient might benefit by the provision of an artificial cushion at the back of the pathway. This can be provided by turning the auricular appendage inside out, like the finger of a glove, passing it through the valve and anchoring the tip inside the ventricle. This manœuvre has the advantage that the cushion is a piece of the heart itself and that it is lined by endocardium. Harken has attempted to achieve the same end by bolstering the postero-lateral cusp away from the wall of the ventricle with plastic balls fixed below it; and other surgeons have introduced a plastic device into the valve which consisted of a ball inside a specially constructed plastic tube.

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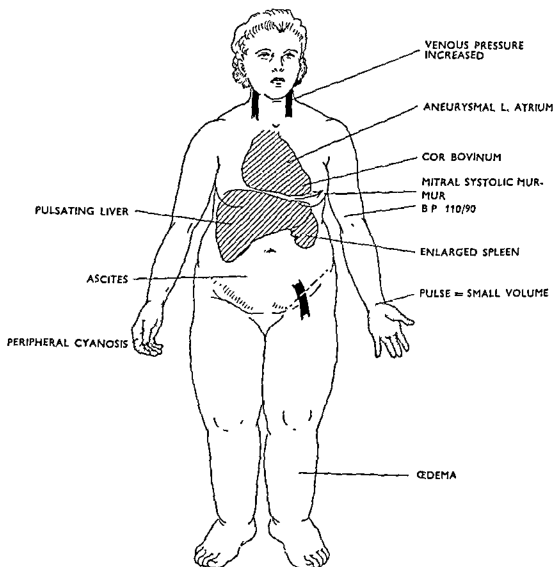


FIG 316 Mitral incompetence

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whole of the heart and great vessels. The technique of splitting the commissures is the same as for other valves, except that some prefer to enter the right atrium by going through its wall rather than through the appendage.

AORTIC STENOSIS

General Remarks. The aortic valve is formed of three cusps which balloon out in diastole and prevent regurgitation by apposition of their margins. Accurate apposition is achieved because the corpora arantii are thrown together in the middle of the aperture. There are no cordæ tendineæ, and the anatomy of the valve is simpler than that of the mitral. More than 400 years ago Leonardo da Vinci showed that the pathway between the aortic cusps was triangular in shape so that its functional size could not be computed by measuring the aperture of the aortic ring, or by passing fingers through the valve in the dead heart.

The surgery of the mitral valve had proved to be so successful that surgeons hoped that the operations upon the aortic valve would be at least as beneficial. In practice this has not been the case, and for the following reasons. The aortic valve is more difficult of access; the diseases, which render it abnormal, also involves the aorta, the adjacent ventricle, the coronary arteries and perhaps other valves; the myocardium is frequently hypertrophied or dilated and it may hold stitches badly; the ventricle is irritable and liable to serious arrythmias during surgical operations; and there are many cases in which no treatment except the provision of an entirely new valve could be of avail.

Pathological Anatomy. Aortic stenosis may be congenital or acquired. The congenital forms are two, valvular stenosis which is probably the result of a fœtal endocarditis in which the ductus arteriosus may remain patent, and subvalvular (subaortic) stenosis probably the result of a developmental defect of torsion of the bulbus arteriosus, in which there may be an associated high interventricular septal defect or a hypoplastic aorta. In either of these varieties the aortic valve may be bicuspid.

The acquired forms may be due to rheumatic infection or to atherosclerotic changes in the valves. In spite of the very different age incidence of the rheumatic and atherosclerotic forms (the former in the first half of life, the latter commonly presenting in the middle sixties) some authors hold that both are manifestations of rheumatic infection.

The pathological appearances of these two forms are different, in the rheumatic the valve cusps are fused at the commissures, are thick, and rolled at the free margins with calcification in the substance of the cusps; the whole forming a funnel-shaped orifice. In the atherosclerotic form the characteristic lesion is masses of calcareous material in the sinuses of Valsalva and only partial adhesion of the commissures. The deposits of calcium may be so dense that opening the commissures can be absolutely impossible even at autopsy. Atherosclerotic valves are not infrequently bicuspid and it has been suggested that bicuspid valves are more liable to degenerative changes as a result of abnormal stress. Bicuspid valves may be a developmental defect or an acquired fusion. Very rarely a rheumatic infection may cause the valve cusp to become retracted in such a way that a subaortic obstruction results.

The mechanical results of obstruction to the outflow of blood from the left ventricle are left ventricular hypertrophy, and post stenotic dilatation, caused by eddies, of the postvalvular portion of the ascending aorta. The actual condition of the myocardium at the time of operation is important to the surgeon.

This artificial valve was introduced into the defect and sewn in place with suture through the myocardium.

Bailey has devised an ingenious technique for suturing the edges of the mitral valve together without opening the heart. The sutures of fascia, or tape, are passed in through the ventricle, up through one edge of the mitral valve, out through the auricular appendage, and then back again through the other edge of the aperture and the ventricle. This leaves a loop at the auricular appendage which can be tightened down, thus approximating the edges of the defect in the valve.

The interest of all these operations is that they show the way in which surgeons are thinking.

TRICUSPID STENOSIS

Stenosis of the tricuspid valve is more rare than mitral stenosis: Paul Wood (1954) has given the following figures for rheumatic heart disease: mitral 80 per cent; aortic 48 per cent; tricuspid 12 per cent; pulmonary 5 per cent.

Diagnosis. Certain points in the history may be noted. A patient who has repeated attacks of œdema and ascites, who can, with the aid of diuretics and rest, lead a reasonably active life may have tricuspid stenosis. Secondly, a patient who has had rheumatic carditis and ascites and who does not suffer from pulmonary symptoms is likely to have an abnormality of the tricuspid valve.

On inspection the face is said to show a particular bronze colour, the explanation for which is not clear. The neck veins are distended and pulsate. The characteristic feature of the latter is the presence of giant "A" waves due to increased pulse pressure in the right atrium. These waves are also transmitted downwards to the liver giving presystolic pulsation which is palpable. The timing of this pulsation enables the surgeon to differentiate between tricuspid stenosis and tricuspid incompetence in which the pulse is systolic.

On auscultation a diastolic murmur can sometimes be detected to the left lower border of the sternum but the difficulty in saying that it emanates from tricuspid valve lies in the fact that tricuspid stenosis is frequently associated with mitral stenosis and other valve lesions.

Radiological examination shows right atrial enlargement, with normal pulmonary arteries and lung fields; and in some cases the pericardium may contain fluid and be enlarged on this account. The electrocardiogram is not pathognomonic, but the P wave is increased and pointed in lead II. Auricular fibrillation is said to be present in 55 per cent of cases.

Treatment. In practice the surgeon is likely to have to deal with some other valve, as well as the tricuspid, in most cases. Brock has operated successfully upon the mitral, aortic, and tricuspid valves at the same operation. Chesterman (1955) gives this advice; "If in doubt (as to which valve to tackle first) we prefer to operate upon the mitral valve first so that excessive pulmonary congestion will not take place. However, if the main disability is due to the raised systemic venous pressure as evidenced by marked congestion of the face and head, we prefer to operate on the tricuspid valve first."

If the tricuspid valve alone is abnormal the operation is done through a right lateral incision but if a more extensive procedure is contemplated the sternum must be split transversely and the incision carried well out into both pleural cavities thus exposing the

whole of the heart and great vessels. The technique of splitting the commissures is the same as for other valves, except that some prefer to enter the right atrium by going through its wall rather than through the appendage.

AORTIC STENOSIS

General Remarks. The aortic valve is formed of three cusps which balloon out in diastole and prevent regurgitation by apposition of their margins. Accurate apposition is achieved because the corpora arantii are thrown together in the middle of the aperture. There are no cordæ tendineæ, and the anatomy of the valve is simpler than that of the mitral. More than 400 years ago Leonardo da Vinci showed that the pathway between the aortic cusps was triangular in shape so that its functional size could not be computed by measuring the aperture of the aortic ring, or by passing fingers through the valve in the dead heart.

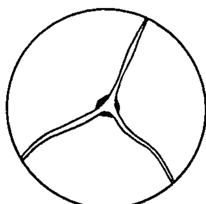
The surgery of the mitral valve had proved to be so successful that surgeons hoped that the operations upon the aortic valve would be at least as beneficial. In practice this has not been the case, and for the following reasons. The aortic valve is more difficult of access; the diseases, which render it abnormal, also involves the aorta, the adjacent ventricle, the coronary arteries and perhaps other valves; the myocardium is frequently hypertrophied or dilated and it may hold stitches badly; the ventricle is irritable and liable to serious arrythmias during surgical operations; and there are many cases in which no treatment except the provision of an entirely new valve could be of avail.

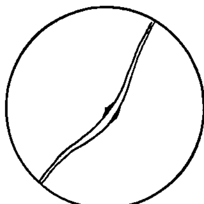
Pathological Anatomy. Aortic stenosis may be congenital or acquired. The congenital forms are two, valvular stenosis which is probably the result of a fœtal endocarditis in which the ductus arteriosus may remain patent, and subvalvular (subaortic) stenosis probably the result of a developmental defect of torsion of the bulbus arteriosus, in which there may be an associated high interventricular septal defect or a hypoplastic aorta. In either of these varieties the aortic valve may be bicuspid.

The acquired forms may be due to rheumatic infection or to atherosclerotic changes in the valves. In spite of the very different age incidence of the rheumatic and atherosclerotic forms (the former in the first half of life, the latter commonly presenting in the middle sixties) some authors hold that both are manifestations of rheumatic infection.

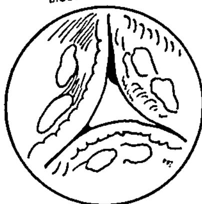
The pathological appearances of these two forms are different, in the rheumatic the valve cusps are fused at the commissures, are thick, and rolled at the free margins with calcification in the substance of the cusps; the whole forming a funnel-shaped orifice. In the atherosclerotic form the characteristic lesion is masses of calcareous material in the sinuses of Valsalva and only partial adhesion of the commissures. The deposits of calcium may be so dense that opening the commissures can be absolutely impossible even at autopsy. Atherosclerotic valves are not infrequently bicuspid and it has been suggested that bicuspid valves are more liable to degenerative changes as a result of abnormal stress. Bicuspid valves may be a developmental defect or an acquired fusion. Very rarely a rheumatic infection may cause the valve cusp to become retracted in such a way that a subaortic obstruction results.

The mechanical results of obstruction to the outflow of blood from the left ventricle are left ventricular hypertrophy, and post stenotic dilatation, caused by eddies, of the postvalvular portion of the ascending aorta. The actual condition of the myocardium at the time of operation is important to the surgeon.



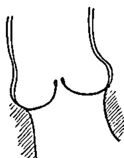


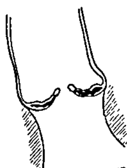


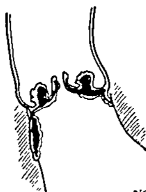


Atherosclerotic valve stenosis. NOTE. Calcareous masses
Rheumatic valve stenosis. NOTE Adhesion along commissures. Less massive calcification

FIG. 317 Aortic valves (seen from above)



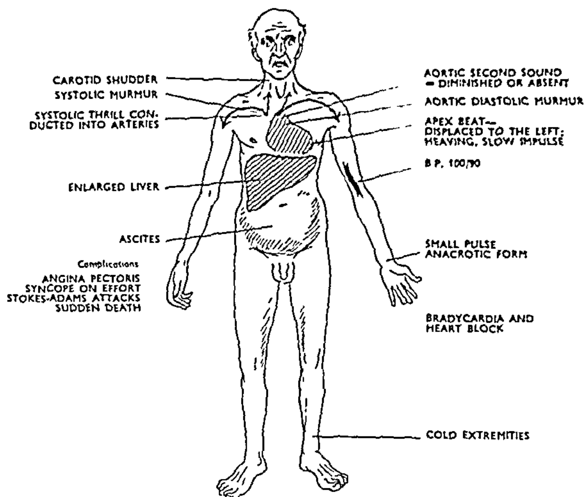




Atherosclerotic valve stenosis NOTE Calcification in myocardium and endocardium adjacent to valve
Subaortic stenosis NOTE Poststenotic dilatation of aorta

FIG. 318 Aortic valves (seen in profile)

In the early stages of aortic stenosis enlargement of the left ventricle may not be very obvious because the hypertrophy of the ventricular wall which occurs encroaches on the ventricular chamber (concentric hypertrophy), for this reason it may be difficult for the surgeon who is inexperienced in these matters, to find the outflow chamber and so to direct an instrument into the valve. When the heart eventually fails the left ventricle dilates, becomes thin and of poor consistency. It not only holds stitches badly but is liable to ventricular fibrillation when manipulated, and to tear when sewn.



PROGNOSIS. Atherosclerotic variety, 60–70 years. Congenital variety, uncertain. Rheumatic variety—depends on other valve lesions. In all cases prognosis is bad when serious *symptoms* appear (i.e. less than 2 years).

FIG. 319. Aortic stenosis.

The Natural History of the Disease. A patient suffering *from aortic* stenosis may live for a long time without much harm or limitation. On the other hand such a one is liable to sudden death at any time. When the heart begins *to fail*, and this moment cannot be predicted, [illegible] There is no [illegible] ortic stenosis [illegible] the patient has started to go downhill.

In the congenital variety there are commonly no symptoms, and the real problem in such patients who are found to have aortic stenosis is the uncertain prognosis. Young patients with the most obvious signs of aortic *stenosis* may be virtually normal. But when

serious symptoms have developed the left ventricular myocardium is unhealthy and operation is dangerous.

The atherosclerotic variety is well tolerated in many cases but once heart failure appears death follows in a few months. Sudden death is not rare and it has been suggested that the valve cusps may become hooked together and obstruct outflow. Incompetence of the valve is often present but it is of much less importance than the stenosis.

Diagnosis. The symptoms are those due to reduced cardiac output, pallor due to peripheral vasoconstriction, syncope on effort due to diminished output on effort (effort *syncope is rare in other forms of heart disease), dyspnœa on exertion, palpitations and a* sense of substernal oppression, and angina pectoris.

The signs are hypertrophy of the left ventricle, a harsh systolic murmur and a thrill which is maximal in the second right interspace and which is conducted in the carotid and subclavian arteries, a carotid systolic shudder or coarse vibration, a diminished or absent aortic second sound. A pulse which is characteristically small and slow rising and anacrotic in form The blood pressure is often low with a small pulse pressure, e.g. 105/95 mm. of mercury. At times the murmur is loudest near the apex beat and then the presence of the murmur in the arteries is a valuable diagnostic point. When the heart fails the murmur may disappear or become uncharacteristic and faint because the cardiac output falls so low that there is insufficient blood flow through the valve to cause it to vibrate forcibly.

On the screen the valves may be seen to be calcified, the left ventricular pulsation is slow and the ascending aorta is dilated. It has been shown that the aortic opening must be reduced to a quarter of the normal before the cardiac output falls.

There is sometimes sinus bradycardia with aortic stenosis; the origin of this is obscure. With the low cardiac output there is coronary insufficiency, and the tachycardia caused by effort further reduces cardiac output and causes myocardial and cerebral ischæmia.

Rheumatic aortic stenosis usually coexists with mitral valve disease and myocardial damage from the rheumatic process, and the prognosis is worse than in the atherosclerotic form. It may be taken as a general rule that the more valves that are involved the worse the prognosis (tricuspid with mitral disease is an exception) and also where muscle and valve disease coexist the prognosis is worse than with pure valve disease.

Selection of Patients for Surgical Treatment. Broadly speaking these cases may be divided into two groups. Those over 50 years of age, and of these it may be said the usual treatment should be conservative for these reasons. Many are symptomless and the signs are picked up on routine chest examination; the average age of death is nearer 70 than 60 (unless important symptoms have developed in which case the expectation is less than 2 years); the valves are usually of the calcific type and are often not amenable to surgical correction. Below the age of 50 the indications for surgical treatment are clearer: they are the presence of progressive symptoms due to aortic stenosis. If, in addition, a significant pressure gradient can be shown to exist across the aortic valve then operation is indicated.

Major Contraindications to Surgical Treatment. *Patients who have reached the* terminal stages of cardiac failure are unlikely to be saved by operation. Angina pectoris is not a contraindication; but definite signs (electrocardiographic) of recent myocardial infarction demand a delay of at least a few months. Some patients have signs of acute rheumatic fever and experience shows that they respond badly to operation: the same is

true if bacterial endocarditis is present. If the heart is very large it is reasonable to infer that the myocardium is exhausted. Massive calcification, as seen on X-rays, does not preclude success, but is not a favourable sign.

Principles of Surgical Treatment and Techniques. The earliest attempts to correct aortic stenosis in Great Britain were made from the distal side of the valve by Brock (1946). These failed not only because an accurate view of the cusps could not be had with the cardioscope he used, but because forcible attempts at commissurotomy with such an instrument were found to be highly dangerous. These early operations were followed by others done through the left ventricle, and in this work Bailey has been a pioneer. The heart is approached by opening the left chest widely and exposing the left ventricle. Before actually deciding that a valvotomy should be done it is wise to measure the gradient of pressures across the diseased valve and these figures can be had by passing a needle, connected to a suitable electrical recording system, directly into the left ventricle and then on into the aorta, or by passing a bronchoscope and introducing a catheter through a needle into the left side of the heart. The difficulty in giving absolute advice on this point is that there is no general agreement of the interpretation of the pressure curves obtained, because they vary according to the type of electrical recorder used. If the patient happens to be suffering from mitral stenosis as well, a much lower gradient will indicate the necessity to operate because in these cases the amount of blood reaching the left ventricle is less. Having decided to do a valvotomy, a spot on the left ventricle, near to the apex of the heart and clear of coronary vessels is selected and a purse-string stitch is inserted into the myocardium. This stitch should be of stout material and placed in a ring of about 1 in. radius. An incision is then made, within the circumference of the purse string, into the left ventricular chamber and bleeding can be controlled by placing a finger on the spot. A valvotome is now inserted. Several different varieties have been devised, and Bailey has produced a special tri-radiate dilator the blades of which are free to rotate. He claims that the expanding blades will naturally fall into position of any remaining commissures and that the instrument is likely to overcome stenosis without creating too much incompetence. Most surgeons prefer a more simple bi-radiate dilator such as has been devised for other valves. The introduction of the instrument into the heart can be difficult for two reasons; if the left ventricular myocardium is hypertrophied the actual chamber may be elusive to find, and if the ventricle is dilated it holds stitches badly, and dangerous bleeding can occur if the purse-string stitch cuts through the muscle. Having got the dilator into the ventricle the closed blades are passed upwards and backwards and can be felt to engage in the valve by palpation of the aortic root from without. If the dilator cannot be passed through the stenosed orifice it can sometimes be guided to the aperture by threading it over a wire guide, as advocated by Bailey. The commissures are separated by forcibly opening the blades of the instrument. From what has been said already it will be apparent that the successful cases will be those in whom an easy split is obtained; the valves which have to be severely forced open are not likely to yield good results as regards subsequent function. This operation is liable at any moment to be complicated by cardiac arrest or ventricular fibrillation. If the heart ceases to pump efficiently before the valvotomy has been done it is probably wise to complete the operation on the valve as quickly as possible and then to apply the routine measures to resuscitate the heart beat and the circulation. Experience has shown that it is most difficult to treat cardiac arrest or fibrillation in the

presence of a stenosed valve. When the valvotome is extracted from the heart the incision is closed by two or three deeply placed interrupted sutures.

When the trans-ventricular operation was first performed the operative mortality was high—Bailey quotes the figure 28 per cent—and the clinical results were disappointing. Increasing surgical experience showed that, even though the details of surgical technique could be refined and improved the risks of this type of operation remained high. With the passage of time the ventricular approach is gradually losing favour and many restrict its use to the following types of case; patients suffering from congenital aortic stenosis or subvalvular stenosis, and those who have a combination of mitral and aortic stenosis. In the latter group the operative mortality has been lower than that for pure aortic stenosis because the obstruction at the mitral valve protects the left ventricle from undue strain. Bailey gives the relative figures as 28 per cent and 18 per cent; he states that in pure aortic stenosis the left ventricle bears the full brunt of the disease. It dilates, degenerates and becomes unduly irritable—all of which load the dice against successful surgery.

For these reasons Bailey advocated a return to the aortic approach from above the valve. He devised the following technique. The ascending aorta was clamped with a lateral clamp which did not occlude the flow of blood through the main channel. The excluded segment of aorta was opened and to this a pouch of pericardium was sewn. Through this pouch a finger could be introduced and in this way the valve could be palpated. Bailey found that the commissures could be separated more often in this way than through the left ventricle; moreover digital separation could be achieved in at least half the cases. In the remainder a suitable knife could be passed in alongside the finger. In 1956 Bailey stated that the mortality for patients operated upon by the trans-aortic route was 14 per cent.

Working on similar lines a number of surgeons have devised different ways of approaching the aortic valve from above. Some have felt that the sewing on of an artificial appendage to the side of the aorta is difficult and dangerous. They have opened the ascending aorta—having arranged hypothermia or a heart-lung machine and clamped off the venæ cavæ, the pulmonary artery and the pulmonary veins—and have then divided the commissures under direct vision. When the aorta has been opened the heart continues to beat, but cannot pump blood, and contrary to expectation the blood in the coronary arteries appears to remain relatively stationary so that air bubbles are not sucked in. When the obstruction at the valve has been overcome the aorta is filled with blood or saline, the clamps are removed from the pulmonary veins, the left ventricle begins to fill with blood, the lateral clamp is reapplied to the aorta, and the circulation is re-established by releasing the cavæ and the pulmonary artery. It is claimed not only that this open approach is the most accurate yet devised but, that the risk of producing dangerous acute incompetence is less than when blind techniques are used.

Yet another solution has been suggested by Sarnoff. He has successfully by-passed the obstructed aortic valve by anastomosing the apex of the left ventricle to the descending thoracic aorta. This operation has succeeded in dogs, but has not been performed in human subjects.

Results of Surgical Treatment. The surgery of the aortic valve is still in the experimental stage and it is too early to assess the results of these surgical endeavours. Nevertheless it is known that the valve can be approached with increasing safety in a variety

of ways, and that an important percentage of the patients can be cured or relieved of their symptoms.

AORTIC INCOMPETENCE

Syphilitic aortitis may cause gross aortic incompetence. In a small proportion of patients with rheumatic heart disease the major lesion is of the aortic valve, the result of which is incompetence. The left ventricle is placed under increasing strain and ultimately

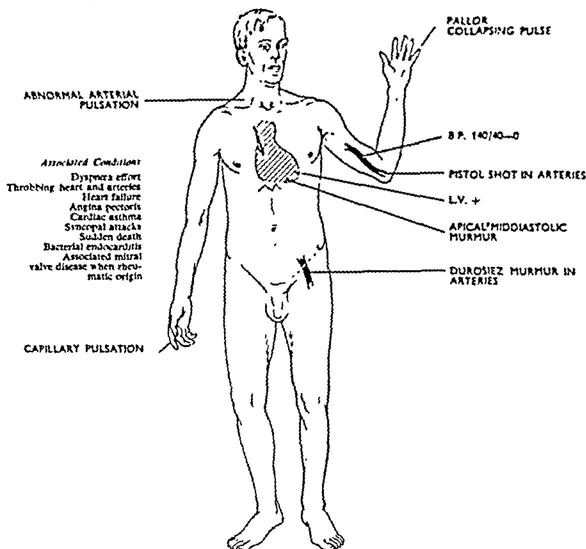


FIG 320 Aortic incompetence.

fails. The throbbing of the arteries both central and peripheral, and the tumultuous heart action, may be very [illegible] either on effort or [illegible] more than a year or [illegible] heart failure occurs the outlook is extremely bad, often only a few weeks or months.

Operations for Aortic Incompetence. A measure of incompetence is present in most patients who have aortic stenosis; but in some the incompetence is the predominant lesion. The problems presented by aortic incompetence are more formidable than those

of aortic stenosis and, as yet, no success has been claimed for direct operations upon the valve itself. Brock (1954) has inserted flaps of pericardium across the aorta above the valve, hoping that, during diastole, the tissue would balloon downwards and restrict the leak. This and other similar measures have failed. Bailey has tried the effect of passing circumferential sutures around the origin of the aorta, but has not succeeded in narrowing the aortic ring without obstructing one or both coronary arteries. Hufnagel (1955) has attacked the problem from another angle. He has devised a plastic ball valve which he puts into the descending aorta below the left subclavian artery. The aorta is cut across and the ball valve inserted between the two ends; it is held in place by two specially constructed plastic cuffs using the principle of multiple point fixation. This does not solve the problem of aortic incompetence but it goes some way to relieving the strain on the left ventricle, because, although the insertion of such a ball valve has no direct effect on the aortic valve incompetence, it reduces the cardiac output by diminishing the rate and quantity of peripheral blood flow in the lower parts of the body. The operation has been done in a number of patients with good results, but practically all cases chosen for surgery have been in advanced heart failure and have been desperate risks. Indeed a number died in the wards whilst awaiting surgical treatment and others were taken to the theatre on several occasions before anæsthesia could be induced. Hufnagel has claimed an operative mortality of 20 per cent and a further 20 per cent have died post-operatively. Many of the survivors are following a useful occupation. In the successful cases the ball of the valve can be distinctly heard clicking up and down, with each heart beat, and this clicking is loud if the patient leans back against a chair or opens the mouth.

ISCHÆMIC HEART DISEASE AND ANGINA PECTORIS

For many years surgeons have appreciated the desirability of treating progressive vascular failure in the extremities; but it is only in recent times that the same ideas have been applied to the heart.

Cardiac ischæmia means anæmia of the myocardium; it is generally produced by coronary arterio-sclerosis; it generally results in progressive diminution of the blood supply to a part of the myocardium with a gradual replacement fibrosis, or it may culminate in pain on effort, or myocardial insufficiency with heart failure, or in arterial thrombosis and myocardial infarction. This sequence of pathological changes might, theoretically, be alleviated surgically in one of three ways: a failing circulation might be improved, pain might be removed by increasing myocardial blood supply, or symptomatic relief of pain might produce symptomatic benefit.

Coronary atherosclerosis is an increasing disease in an ageing population; it is responsible for about a quarter of organic heart illness and is the cause of 80 per cent of sudden deaths. It affects men about four times more commonly than women, and has its highest incidence amongst professional classes. The cause is not known.

The most dramatic symptom of myocardial anoxia is angina pectoris. The term angina pectoris is applied to attacks of chest pain, of short duration caused by effort or excitement usually consisting of a sense of ache or oppression or constriction beneath the sternum, which may radiate to the arms or neck or jaw. It is usually produced by exertion or emotion and relieved by rest. Occasionally it occurs, in patients lying in bed, with change in position. It is a symptom and may occur without any other detectable abnormality, including a normal E.C.G. and normal chest X-rays. Angina pectoris does not

occur in every patient who has coronary atherosclerosis. It may be due to other causes such as syphilitic aortitis, valvular disease (particularly aortic stenosis), paroxysmal tachycardia, etc. Anæmia, obesity, gall bladder and gastro-œsophageal disease may be contributory factors.

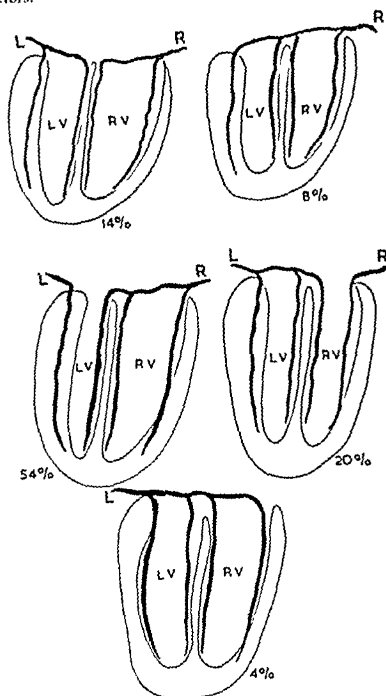


FIG 321 Coronary artery, distribution on the posterior surface of the heart (after Barnes).

Surgical Anatomy. There are two coronary arteries which arise from the aortic sinus immediately distal to the aortic valve. *The left* comes forward between the root of the pulmonary artery and the left atrium and divides into an anterior descending branch and a left circumflex: the former runs down the front of the heart, from the

of aortic stenosis and, as yet, no success has been claimed for direct operations upon the valve itself. Brock (1954) has inserted flaps of pericardium across the aorta above the valve, hoping that, during diastole, the tissue would balloon downwards and restrict the leak. This and other similar measures have failed. Bailey has tried the effect of passing circumferential sutures around the origin of the aorta, but has not succeeded in narrowing the aortic ring without obstructing one or both coronary arteries. Hufnagel (1955) has attacked the problem from another angle. He has devised a plastic ball valve which he puts into the descending aorta below the left subclavian artery. The aorta is cut across and the ball valve inserted between the two ends; it is held in place by two specially constructed plastic cuffs using the principle of multiple point fixation. This does not solve the problem of aortic incompetence but it goes some way to relieving the strain on the left ventricle, because, although the insertion of such a ball valve has no direct effect on the aortic valve incompetence, it reduces the cardiac output by diminishing the rate and quantity of peripheral blood flow in the lower parts of the body. The operation has been done in a number of patients with good results, but practically all cases chosen for surgery have been in advanced heart failure and have been desperate risks. Indeed a number died in the wards whilst awaiting surgical treatment and others were taken to the theatre on several occasions before anæsthesia could be induced. Hufnagel has claimed an operative mortality of 20 per cent and a further 20 per cent have died post-operatively. Many of the survivors are following a useful occupation. In the successful cases the *ball of the valve can be distinctly heard clicking up and down, with each heart beat,* and this clicking is loud if the patient leans back against a chair or opens the mouth.

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has been divided at operation, without obvious harmful effect. On the other hand maintenance of the coronary circulation is at least as vital as keeping the brain alive: the one is useless without the other.

In atherosclerosis the changes which ultimately destroy the heart are imposed slowly. They start in the walls of the coronary arteries and resemble atheroma. Lipoid substances accumulate in the intima, and the media is progressively replaced by fibrous tissue. Calcification and deposits of bone occur in the walls of the arteries converting them into rigid tubes which, although anatomically applied to the surface of the ventricles, cannot move in concert with them. The diseased arteries are prone to thrombosis and to sudden occlusion from embolus, subintimal hæmorrhage, etc.

The result of these changes is that the coronary vessels become progressively incapable of supplying the circulatory demands of the myocardium. In some patients the effect is that a diffuse fibrosis of the heart occurs, and in others a sudden episode of arterial occlusion precipitates a myocardial infarct. But in some patients the heart muscle remains normal even though the coronary arteries are occluded and in these myocardial ischæmia with angina pectoris may occur with effort.

An infarct can be distinguished with the naked eye within 24 hours although microscopic changes of cell death are present sooner than this. At this time the affected area of myocardium moves paradoxically as the heart beats; but an infarct seldom extends right through the myocardium from surface to surface: if it did, rupture of the heart and cardiac aneurysm would be more common. Within a few days the infarcted area becomes œdematous and surrounded by a zone of hyperæmia. It is ultimately replaced by scar, and no longer acts as part of the cardiac pump.

The surgeon might intervene in this sequence of events at several points. He might recognize progressive diminution in the myocardial circulation, and attempt to augment it before fatal damage had occurred to any part: he might improve the circulation of the remainder after an infarct had destroyed a part: he might alleviate some of the symptoms of angina pectoris or he might excise and repair a cardiac aneurysm.

Beck's View of Myocardial Infarction. Beck has produced myocardial infarction experimentally and he has described the events as follows:

"The fatal heart attack is similar to a convulsive seizure. It is now recognized that an area of anoxæmia in the brain gives rise to a convulsion. This focus on the brain has been referred to as a trigger. The trigger discharges and from it emanate impulses to other parts of the brain. These produce a generalized convulsion. An analogous condition exists in the heart. A trigger can be made by ligation of four or five coronary channels supplying an area of heart muscle 2 or 3 cm. in diameter. This area of myocardium becomes cyanotic after these arteries are ligated and suddenly something happens which destroys the co-ordinated heartbeat. Some fibres are contracted while others are elongated and the ventricles become a writhing squirming mass of muscle which does not pump out any blood. The term ventricular fibrillation is applied to this condition and, as you know, it is fatal. It is similar to a *convulsion* of skeletal muscle and the term convulsion of the heart might well be applied to this condition. I believe if we could actually observe the heart in fatal attacks we would prefer to use the term convulsion of the heart rather than ventricular fibrillation.

"I should like to return to the experiment in which we ligated arterial channels to produce a trigger. I should like you to visualize an experiment in which we ligate four

auriculoventricular groove to the apex, and is a fairly accurate surface marking of the interventricular septum. This artery supplies the front of the right ventricle, the interventricular septum, the front and the top of the left ventricle. The circumflex branch runs between the left atrium and the left ventricle to the back of the heart and supplies the upper part, the back and sides of the left ventricle.

The right coronary artery has no large division: it passes to the right between the right atrium and the right ventricle and supplies part of the front of the right ventricle and the back of both ventricles. There is some variation in the supply of the backs of the ventricles and these variations are shown in the diagram (Fig. 321).

At all ages in normal persons only minor anastomotic channels exist between the main coronary arteries and their branches, but intercapillary connections are common. When the coronary arteries are diseased the number of anastomoses increases and in such cases anastomotic channels between the bronchial arteries and the coronary arteries open through the vessels which are the vasa vasora of the first parts of the aorta and the pulmonary artery. Anastomoses also occur when the pericardial layers become adherent, but whether these naturally occurring pericardial anastomoses, or those which form following deliberate pericardial injury, actually carry blood into the myocardium is controversial.

The most secure part of the heart in blood supply is the top of the left ventricle; the least safe is the apex, the back and side of the left ventricle.

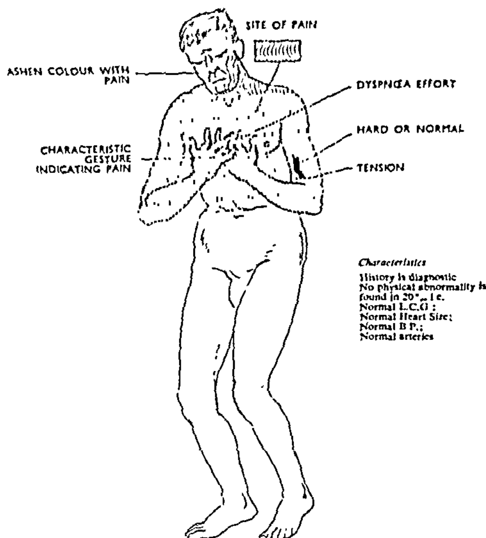
Any part of the heart may be affected by coronary disease but thrombosis involves the anterior descending branch of the left coronary artery in about 50 per cent of cases, and the right main coronary in about 23 per cent. The left circumflex branch is involved in 18 per cent. The left main coronary artery in 4 per cent. In most patients coronary disease is not limited to one zone, but the common sites of infarction correspond with the above figures. Most of the blood returning from the myocardium drains into the *coronary sinus*. This is a large venous channel, about 5 cm. long, which lies half embedded in fat in the back of the auriculo-ventricular groove. It runs across the back of the heart from left to right: at its beginning it is in fairly close proximity to the thoracic aorta, the pericardium intervening, and at its termination it opens into the right atrium close to the inferior vena cava. In life this sinus is distended and obvious: in the dissecting room it escapes detection because it is collapsed. The orifice of the coronary sinus is guarded by the valve of Thebesius. The sinus is joined by five fairly constant veins and the orifices of all these into the sinus are said by Gray to be guarded by valves: this point is important to the surgeon who may consider reversing the flow of blood in the sinus (q.v.). These veins are the great, small, and middle cardiac veins, the posterior vein of the left ventricle and the oblique vein of the left atrium. Apart from all these vessels there are numerous other channels which drain venous blood directly into the right and the left heart chambers.

To the surgeon who wishes to augment the blood supply to any part of the heart, the coronary arteries, which are diseased, could be of less interest than the veins which are normal.

Surgical Pathology. The effects of interrupting the blood supply to various parts of the heart in man depends upon the normality or otherwise of the coronary arteries at the time. For instances cases are on record of complete division by trauma of the anterior descending branch of the left coronary artery, in young patients who have not developed infarction: in some patients suffering from Pick's disease a branch of the coronary artery

Angina Pectoris

The diagnosis of angina pectoris rests upon the existence of a variable chest pain. It is mid sternal and sometimes excruciating: it tends to radiate to both arms and perhaps to the neck and the jaw; it may pass through the thorax to the back. It is very rarely localized to the left mammary area. It is described as constricting, squeezing, or



Prognosis Variable spontaneous disappearance of pain 5-10%.

Fig 322 Angina pectoris.

bursting, and whilst it lasts it is continuous and without fluctuation. It is not sharp, *stabbing*, or *paroxysmal*. An attack does not last more than a few minutes and it is provoked by an exertion or emotion which imposes extra work upon the myocardium particularly after a meal and *in cold weather*. The diagnosis is made on taking the history: there may be no supporting physical signs on which the physician may be able to determine the cause of the attack. *Relief obtained* by taking nitrites is a diagnostic feature.

The Prognosis of myocardial infarction ... in different individuals; Fe

small channels to a small area of myocardium. We can observe the cyanosis of this small area of myocardium. Now if we go no farther with our ligation we can close the operative wound and the animal will recover. If we go ahead and ligate the fifth channel to this small area of myocardium the trigger will discharge, destroy the co-ordinated beat, and produce death. This fifth small channel then can be regarded as the 'last straw.' It makes the difference between life and death. This vessel carries a small amount of blood, and a small amount of blood can make the difference between life and death. This small amount of blood can be looked upon as a blood-bath. I do not think we can over-emphasize the importance of a small amount of oxygenated blood delivered to that part of the heart where the blood supply is deficient. Those who are adversely critical of this research will please bear in mind that a small amount of oxygenated blood can be significant. It can save the life of the individual who has a trigger zone developing in the heart. I need scarcely remind you that when death occurs from a heart attack it is usually sudden and unexpected. Usually there are no signs of heart-failure. This is a physiological derangement and it is as effective as turning the ignition switch in a smoothly working motor. The trigger zone may be important anatomically. No doubt it could be excised experimentally if one cared to do so and the animal would be left with a good heart. A new set of coronary arteries is not necessary to prevent a small localized trigger from discharging. A blood-bath can be effective."

The Diagnosis of Ischæmic Heart Disease will be briefly considered.

Myocardial Infarction

The history is of great importance. The age, the sex, the mentality, and occupation of the patient; the presence of previous attacks and other conditions, such as hypertension, syphilis, etc., are suggestive of the diagnosis. The onset of infarction may be symptomless, it may cause angina pectoris only, or lead to slight pain with or without shock. On the other hand in many cases it is sudden and overwhelming; it often occurs irrespective of the activity of the patient. The first symptom is usually severe pain, of anginal type and distribution, but lasting for a long time. The important featûre is that the pain is continuous over a period of time, usually more than a few minutes.

The intensity of the pain does not correspond to the size of the infarct; it varies from mild to intolerable. Morphia alone deadens it. The signs are those of shock, dyspnœa, and perhaps loss of consciousness. The patient is generally frightened and dyspnœic. The pulse is rapid and feeble; the peripheral circulation is poor. The blood pressure does not always fall immediately but ultimately drops, and can be low for weeks afterwards. In very severe cases the heart sounds are weak and sometimes pericardial friction occurs. Disturbances of rhythm occur.

The electrocardiogram can be normal after myocardial infarction but, in 75 per cent of cases if full chest lead studies are made, it provides an accurate proof of the presence and position of the infarct. The radiological examinations are not practicable in the acute phase but may help in the diagnosis of complications.

The acute phase of the illness may be fatal but if the patient survives, the symptoms take 2–3 days to subside unless there is cardiac failure, further infarction or embolism. Ventricular fibrillation, left ventricular failure, pulmonary embolism, peripheral venous thromboses and systemic emboli are the dangerous complications.

Blumgart and others advocated total thyroidectomy. These operations have been discarded, and if necessary comparable results might be achieved by exhibiting thiouracil.

Operations have been devised *to increase the blood supply to the heart* by grafting omentum, substernal fat, the internal mammary artery, the lung, or the pericardium to its surface. Beck was the first to show that it was possible to provide a collateral circulation in this way and that such a circulation carried a lot of extra blood to the myocardium; moreover he proved that it could prevent the "trigger mechanism," described above, from discharging. It has been shown that new vessels grow into the myocardium, and that they can react to inflammation or ward off the serious effects of ischæmia if a main branch of a coronary artery is destroyed.

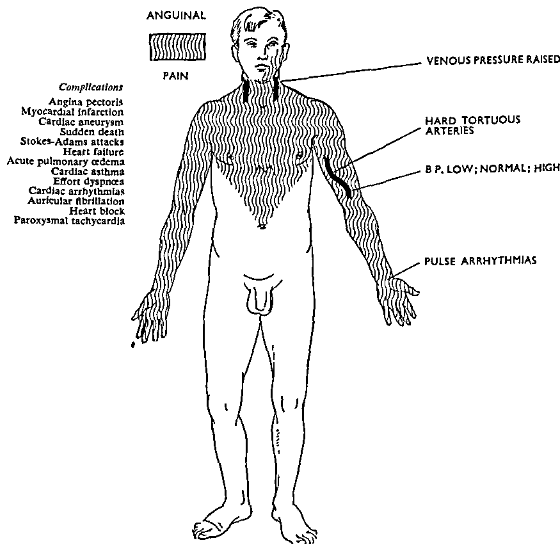
O'Shaughnessy was the first to popularize these operations in England. He used an omental graft and published his results in 1936. This work has never been followed with enthusiasm by other surgeons: one of the technical difficulties about it is to bring the omentum up through the diaphragm without starting a diaphragmatic hernia. The clinical results of these operations have not been adequately investigated owing to the untimely deaths of O'Shaughnessy and Mansell. Beck has preferred producing adhesions between the heart and the pericardium by introducing irritating substances and abrading the surface of the ventricles. Talc, asbestos powder, and aleuronat paste have been recommended, and Mason, in England, following Beck's technique has reported upon these methods. On the whole these operations, although not generally favoured by physicians, seem to be worthwhile. Mason's results were: 30 patients were operated upon, 11 died within a month, 3 were not relieved of their symptoms, but 16 were improved and some of them were much better than before operation.

Murray and other surgeons have devised methods of anastomosing various systemic arteries to the appropriate coronary artery beyond the developing obstruction. Murray stated that in at least 50 per cent of cases obstruction in the left anterior descending coronary artery was of strictly limited extent. His treatment was to mobilize the left subclavian artery, increase its length by grafting a segment of the radial artery to its end, and to join this, end to side, to the coronary artery beyond the obstruction. These operations depend for success on the fact that a freely bleeding artery implanted into the myocardium will not produce a hæmatoma, because the myocardial drainage carries off the blood effused. The internal mammary vessel has been preferred by other Canadian surgeons.

Ingenious attempts to improve the blood supply of the myocardium have been made by *operations upon the coronary sinus* and its tributaries. Gross and others showed that simple ligation of the coronary sinus in the dog caused an increase in the coronary vascular tree; and preliminary preparation of a dog in this way made it impossible to produce infarction by subsequently tying the left descending coronary vessel. Fauteux and Palmer showed that a similar protection was provided by ligation of the vein at the same time as the artery. They applied this, with considerable success, to a group of patients in whom they ligated the great cardiac vein. Fauteux, later, combined this operation with denervation of the heart and stated that the results of the combined procedure were even better. He operated principally to relieve angina pectoris in patients who were totally incapacitated. Patients over the age of 60 were not good operative risks, nor were those who had serious disturbances of rhythm. The operations were done under ether anæsthesia, and the front of the heart was exposed through a left-sided trapdoor incision.

the duration of life was 3·7 years. He pointed out that conventional medical treatment "does not in any way alter the underlying progress of the changes." Palmer stated that 73 per cent of patients can expect to be alive after 4 years and 38 per cent after 10 years. Social and economic factors influence the outlook after infarction.

The prognosis of angina pectoris is extremely variable, some patients die early, others



Prognosis: Very variable. After myocardial infarction 30% live 10 years

FIG. 323. Coronary artery disease

have pain on effort for many years, and still others get a spontaneous remission of pain—this applies particularly when anxiety over some problem is relieved. This extreme variability of outlook makes any assessment of results difficult and any selection of patients for operation equally difficult. However, where anginal pain occurs with such little effort as to prove crippling or when anginal pain occurs at rest or on minor effort some surgical relief can be given.

The Surgical Treatment of Cardiac Ischæmia

The surgical treatments which have been tried are these:

Attempts to *reduce the metabolic requirements* of the body have been made. In 1933

The aim of investigations today is not only to assess the anatomy of aneurysms and to find out which vessels are involved, but to ascertain the ætiology and the normality or otherwise of adjacent structures. In this chapter an aneurysm is regarded as a permanent dilatation of a part of an artery due to pathological changes in the walls of that vessel.

Cardiac Aneurysm

Aneurysm of the Ventricle. Until recently ventricular aneurysm was not distinguished from hypertrophy or dilatation of the heart. The common cause is myocardial infarction of the left ventricle involving the anterior or the posterior wall, and the aneurysm is usually complicated by generalized coronary arterial disease. A rare cause is a heart wound, either due to surgery, to a foreign body, or to a stab. In most survivors heart wounds heal perfectly. It is believed by some that continued activity during convalescence from an episode of myocardial infarction is an important predisposing factor. The aneurysm develops as a blow-out from the wall of the ventricle after the infarction has been partially replaced by scar tissue. The affected area fails to pulsate or it moves paradoxically *vis-à-vis* the rest of the ventricle, that is, it distends during systole, and this paradoxical movement can sometimes be discerned on screening; it is a valuable diagnostic point. The lesion grows as a bulging, pulsating area connected with the wall of the ventricle. As the aneurysm increases in size it forms a cul-de-sac in which thrombi develop and these are apt to separate and to cause dangerous systemic embolism. The neck of the aneurysm is quite likely to remain small and of such a size as to be manageable surgically; the tumour is rather sessile in shape; it remains confined within the pericardium to which it is often adherent, and as it increases in mass, it may cause a pericardial effusion. Calcification of the wall of the aneurysm and its contents may occur.

Diagnosis. There are no special diagnostic criteria; the condition is likely to be picked up during radiological investigation of a patient suffering from myocardial infarction, but a case recently seen by the author occurred in a child who had previously undergone a cardiotomy for pulmonary stenosis. Angiocardiography may prove that an opacity seen on radiographs to be connected with the heart is in fact an aneurysm.

Treatment. The outlook is bad and, untreated, the patients die of coronary insufficiency or congestive heart failure. Rupture of the aneurysm is extremely rare. Some cases will be suitable for surgical treatment, and there are records of successful operations. If the aneurysm has a neck which can be clamped the sac can sometimes be excised and the cut edges of the myocardium approximated with sutures. The factors which limit surgical intervention are generalized coronary disease and a broad neck to the tumour. Bailey has operated successfully in this condition.

Congenital Aneurysms. Chiefly affect the atria. Very few have been diagnosed in the past, but angiocardiography should correct this. Sauerbruch operated upon two patients prior to 1939. They were thought pre-operatively to be suffering from mediastinal tumours. Sauerbruch exposed the lesions, clamped off the bases, excised the tumours and sewed up the defects. Both patients did well. Other successful excisions have been described recently.

Arterio-venous Aneurysm occurs between the pulmonary artery and a branch of the left coronary artery. Crafoord reported such a case which he saw in a child who was

The great cardiac vein was exposed and a ligature passed round it, but not tied. The heart was denervated by removing the adventitia from the ascending aorta and the pulmonary artery, and, when this had been done, the great vein was tied off.

Beck has tried to *revascularize the heart directly from the aorta.* To do this he removed a length of one of the systemic veins or arteries, such as the brachial; anastomosed one end of the graft to the descending aorta behind the pericardium, and joined the other end to the coronary sinus on the back of the heart. The coronary sinus was partially obstructed at its entrance into the right atrium at the first operation and subsequently was tied off completely. The effect of this procedure was to take blood straight from the aorta into the venous system of the heart and to rely upon the Thebesian veins for its return to the circulation. The operation has been successfully performed in human beings and clinical improvement has been claimed. Bailey, who has carried out many experiments on these lines, finds that the myocardium can be revascularized by operations of this type but that, in animals, the anastomoses close after a few months.

The pain of cardiac ischæmia can generally be relieved by *interrupting the nerve pathways to the heart.* The possible benefits which might accrue from this are that angina might be controlled and the "trigger mechanism" might be diminished. On the other hand the pain is produced by anoxia. To relieve the pain might increase the risk of the disease.

Most anatomists agree that the normal pain fibres pass through the third and fourth sympathetic ganglia on both sides: from there they pass through the white rami communicantes and enter the spinal cord through the posterior roots of the corresponding segments. The operations which have been practised are, posterior rhizotomy in which the upper 4 or 5 thoracic roots are divided; paravertebral block with procaine or alcohol, and cervico-thoracic ganglionectomy.

The last is the procedure of choice; the stellate and the upper 3 thoracic ganglia are removed on one or both sides. White and Smithwick approached the ganglion under general anæsthesia, from the front of the neck and they removed the necessary parts by dissecting the mediastinal pleura forwards. Many patients have been treated in this way. Lindgren, working in Sweden, has reviewed the end results in 105 cases: the operative mortality was 8·5 per cent and the remote mortality up to 2 years was the same in patients who underwent operation as in those who did not. Three-quarters of the patients had relief from pain on the side of the operation, and in two-thirds the capacity for work was increased. More than half had a bilateral operation. The Swedish surgeons believe that the warning signal is not removed by operation because all these patients had some discomfort on exercise after treatment, and this they considered to be due to impulses transmitted by the vagus nerves.

THORACIC ANEURYSM

General. One hundred years ago the clinical diagnosis and treatment of aneurysm comprehended an important part of surgical thought and achievement. In the early part of this century the condition was of waning interest. It is now becoming important once again. Two changes have recently affected the diagnosis and treatment. The first is the decline of syphilis, and the relative increase of other diseases as ætiological factors; and the second is that active, as opposed to palliative, surgical measures can often be entertained.

is often dull and described as a feeling of fullness—and repeated small hæmoptyses. The common cause of death is hæmoptysis. Rusby has described some patients in whom no demonstrable change occurred in the tumour over a period of many years, and in these people the general health remained good.

The importance of these cases is that they present as a para-mediastinal, or para-cardial shadow on radiograms and this is often erroneously diagnosed as something else. There are no typical murmurs or electrocardiographic changes due to the aneurysm itself. The mass does not always pulsate on screening or kymography. The diagnosis depends upon angiocardiography which is conclusive; but catheterization may be needed on occasion to exclude an aneurysmal ductus arteriosus.

Treatment. In most cases surgical treatment is unnecessary, but a demonstrably progressive increase in size and continuing hæmoptysis are danger signals which may demand ligation of the relevant pulmonary artery on either side of the swelling. This procedure might have to be associated with pneumonectomy but in most cases the bronchial arteries should be adequate to maintain the life, and some function of the lung. And the possibility of excising the aneurysm and bridging the gap with a graft should be entertained. The ductus arteriosus may be involved in such an aneurysm.

Aneurysm of the Aorta or its Thoracic Branches

Aneurysm of the aorta is still fairly common and, in 1948, about 1,500 persons died from the disease in England and Wales.

Ætiology. Ninety per cent of cases used to be due to acquired syphilis (congenital syphilis is said not to produce aortitis) and many of the patients ultimately developed signs and symptoms of neuro-syphilis as well as aneurysm. In this group the serological reactions for syphilis is usually positive, but in recent times syphilis has declined as a cause, and many examples of arteriosclerotic aneurysm, congenital aneurysm, and aneurysm due to idiopathic hypoplasia of the media have been recognized. The latter have proved more amenable to curative surgery than syphilitic lesions, and for this reason it is now important to establish the ætiology before contemplating treatment.

Surgical Pathology. The characteristic pathological changes were accurately described by Astley Cooper 125 years ago. Three types of aneurysm occur and are called saccular, fusiform, and dissecting. A saccular aneurysm is one in which the tumour is a diverticulum from the main blood pathway, its walls are formed of fibrous tissue and adjacent structures; in fusiform aneurysm the tumour involves the lumen of the aorta itself or one of its branches; and in dissecting aneurysm the blood dissects the medial coat of the vessel concerned. An important difference between aneurysms due to syphilis and those due to other causes is that the walls of the former are dense, adherent, and invasive; whereas the latter are often free from the structures they touch and the walls may be so thin that the blood can be seen circulating within. Syphilitic endarteritis of the vasovasorum and atheroma of the intima occur at first in patches, and result in weak spots which coalesce and spread. The aneurysm which develops in such a vessel has walls consisting of dense scar tissue, which may contain plaques of calcification or bone. Within the sac there are laminated layers of organizing clot, and in some fortunate individuals nature effects a cure by obliterating the diverticulum in this way. As the growing tumour impinges upon adjacent structures they are displaced or destroyed according to their mobility. For instance the trachea, the bronchi and the œsophagus

diagnosed wrongly as suffering from a patent ductus arteriosus, because of the typical machinery murmur which was present. He operated; diagnosed the lesion; divided the communication and cured the patient.

Aneurysm of an Aortic Sinus of Valsalva. This condition was generally thought to be due to syphilis; nowadays it is regarded as being also mycotic or congenital. There are three sinuses and aneurysm may develop from any of them and rarely from more than one. The situation of the aneurysm predisposes to certain complications among which are the following:

(1) Right Sinus of Valsalva.

This may rupture into the superior vena cava or compress it and cause obstruction, or rupture into the right atrium or the right ventricle or more rarely into the left ventricle.

(2) Left Sinus of Valsalva.

This may compress the pulmonary artery or rupture into it.

(3) Posterior Sinus of Valsalva.

This may compress the left main bronchus, or rupture into the left ventricle.

Aortic incompetence and bundle branch block are associated lesions. The tumour seldom grows large but Kerley has described a case in which it impinged upon the left main bronchus and caused pulmonary atelectasis. The patient had been diagnosed, on routine radiology, as having a left-sided mediastinal mass, but the true diagnosis was revealed by angiocardiography. In most cases the aneurysm ruptures sooner or later. If the perforation occurs into the pericardium or into the pleural cavity the patient dies; but if it ruptures into the vena cava, right atrium, right ventricle, or the pulmonary artery a well defined syndrome occurs which may be compatible with life. If the rupture is into the right heart the patient develops signs and symptoms due to overloading of the pulmonary circulation, and cardiac failure follows. On cardiac catheterization, samples of blood resemble those secured in patients suffering from septal defects. Perforation into the pulmonary artery produces the typical signs of a patent ductus arteriosus.

There are, as yet, no accounts of surgical treatment of these cases but the problems presented are not insuperable and the mechanical disabilities should be overcome by ligation or excision of the aneurysm.

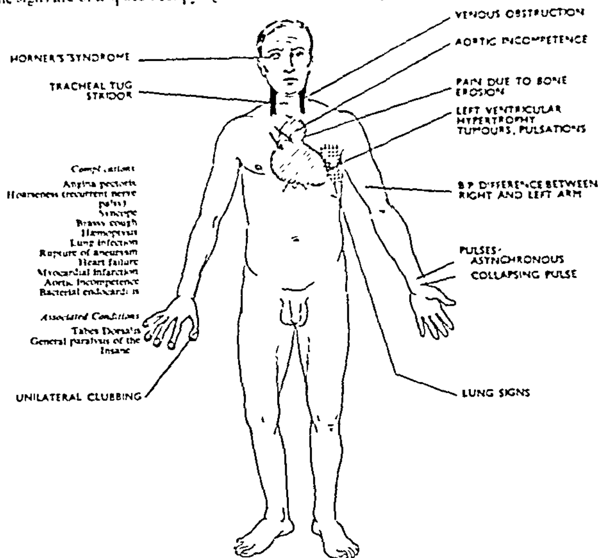
Aneurysm of the Pulmonary Artery

Aneurysm of the pulmonary artery is a *relatively common lesion*, and more than 200 undoubted examples have been described in the literature. The aneurysm usually affects the right or the left main branch and can reach the size of a large orange. The remarks which follow refer to those cases where the lesion is solitary; they do not describe post-stenotic dilatation, dilatations associated with a reversed shunt in a patent ductus arteriosus, or dilatations of the pulmonary artery associated with atrial septal defects.

Ætiology and Anatomy. There are five known ætiological factors. Congenital aneurysm is the commonest (40 per cent). The second factor is syphilis (30 per cent) and in these patients aortic aneurysm may be present as well. Infective arteritis, atheroma and trauma are the other causes, among which schistosomiasis is important in areas where infection is endemic.

Aneurysm of the pulmonary artery occurs in a younger age group than aortic aneurysm and is equally common in the two sexes. It may be silent and only revealed on mass radiography, but the usual symptoms are exertional dyspnœa, cough, pain—which

The symptoms include local pain, hæmoptysis, backache, dyspnœa on exertion, cough, hoarseness, dysphagia, and defects due to vena caval obstruction and pulmonary atelectasis. Any of the mediastinal nerves may be paralysed. Tracheal tug may occur. The signs are of a space occupying mediastinal mass, and palpable pulsation occurs when



Prognosis Variable, 2 years after diagnosis.

FIG. 324 Aortic aneurysm

the aneurysm approaches the skin. Aortic arch aneurysms come to the surface in the suprasternal notch or through the sternum; those of the innominate artery appear to one side of the sternum or in the posterior triangle of the neck. The Wassermann and Kahn reactions are generally positive. Cardiac enlargement occurs with aortic incompetence which can be detected by an early diastolic murmur to the right or left of the sternum, which may be associated with a collapsing pulse and large pulse pressure. Cardiac enlargement may also occur with coronary ostial disease which leads to myocardial insufficiency from ischæmia. Angina pectoris, sudden syncope, and heart failure occur with these complications. The aneurysm itself gives rise to no pathognomonic murmurs and heart rhythm is regular.

are pushed to one side and it is only in the late stages that rupture into these viscera occurs. By contrast limiting bone is completely destroyed and this is well seen in the vertebral column, the sternum, and the ribs. There has never been a satisfactory explanation of why an aneurysm "erodes bone." Nerves, such as the phrenics and the recurrent laryngeal, become included in the wall of the "tumour" and could be paralysed eventually; the "brassy" cough, which is a sign in a few cases, is more often due to pressure on the air passages than to recurrent nerve palsy. As an aneurysm approaches the skin it forms a prominence which increases in size; the subcutaneous tissues thin out and the area becomes red, inflamed, tender, and pulsating. Towards the end the central patch of skin dies and forms a black scab; the pulsations and the pain increase and threatening specks of blood appear on the dressings. Death is nearly always due to rupture of the aneurysm and this may occur, not only through the skin, but into the œsophagus, the air passages, the pericardium or the pleural cavity.

The points which particularly affect the surgeon are these. A high proportion of aortic aneurysms are saccular in type and remain so however large they become. Moreover the original defect in the wall of the aorta often does not increase in size as the "tumour" grows. The whole of the thoracic aorta may be abnormal in these cases, but quite often the area where the aneurysm has blown out is the only really bad part. These aneurysms often occur in patients who have no other signs of active syphilis, and if they could be controlled or eliminated surgically the outlook for the patient would be excellent. A syphilitic aneurysm is not a sharply defined mass whatever it may look like in the radiographs, it is densely adherent in most places, and least adherent at its neck. If the aneurysm is involving bone it cannot be dissected free without opening the lumen unless it contains organized clot in the relevant area. It may include important nerves in the walls. The distal part of the sac is poor material for suture or anastomosis, and if wrapping is contemplated the surgeon should know that he will probably not be able to mobilize the most dangerous area. The size of an aneurysm is often not as vital as its anatomical site; the writer has seen an aneurysm about the size of a plum which protruded from the concave margin of the aortic arch (and may have arisen in a partial occlusion of the ductus in which the aortic end had remained patent) which caused atelectasis of the left lung and which burst into the air passage on bronchoscopy. In some fusiform aneurysms the defect is sharply limited to one reach of the vessels and the remainder is of normal appearance; in others the tumour is elongated, bossed, and of complicated form. Multiple aneurysm occur and the demonstration of one should be a spur to find others before considering surgery. The patient dies eventually of cardiac failure due to aortic incompetence or coronary ostial stenosis. More rarely of the infective complications, of bronchial compression, or from rupture of the aneurysm.

Diagnosis. An aneurysm may be silent but in most cases it is not. The diagnosis depends ultimately upon radiology, angiocardiography, and kymography. Kerley reported a series of more than 50 patients sent to him for radiological examination, in not one of whom was the correct diagnosis made on physical signs in the out-patient department. By the time a thoracic aneurysm is obvious clinically the disease is generally far advanced. The differential diagnosis is from other mediastinal tumours and the commonest mistakes are when an aneurysm is confused with a mediastinal dermoid, or mediastinal goitre, carcinoma of the lung, a mass of enlarged glands, or an "uncoiled aorta."

Prognosis. The prognosis of thoracic aneurysm is bad although some may remain silent for a long time; in some cases the duration of life is less than 2 years after the symptoms are first manifest, and during this time the patients suffer much pain. The cause of death is heart failure, pressure upon, or rupture into, some vital structure. Aneurysms of the transverse aorta are especially dangerous: out of 219 patients collected by Colt one half died in less than 10 months, and only a quarter lived 2 years.

It is generally taught that the cardinal sign of a dissecting aneurysm is excruciating pain; that the accident produces urgent signs and symptoms and an early fatal result. In fact dissecting aneurysms may be seen in which pain is not prominent, the clinical course is benign and healing by fibrosis and calcification occurs. In this group hypoplasia of the media with cystic medionecrosis is the underlying lesion and conditions such as arachnodactyly and dislocation of the lens may be present at the same time.

Treatment of Aortic Aneurysm. The medical treatment of syphilitic aneurysm is directed against the cause of the disease: in addition heart failure and pain can be alleviated to some extent. But the progress and outcome of the aneurysm is not often affected, and the majority of the patients die within a year.

Until recently the surgical treatment has hardly been more satisfactory: it is so often limited by the fact that the patients are old, they have a defective myocardium and heart failure, or are ill with pain and lung infection, or neurological illness.

The safest procedures are those which attempt to increase the clot within the sac, that is, to emulate nature's way of dealing with the problem. The first surgeon to suggest introducing loops of wire into an aneurysm for this purpose was Moore in 1864. On the whole the method was not successful and had virtually been given up when Colt and Power (1903) introduced their wire wisps for this purpose. The operation is only practicable when the aneurysm is adherent to the chest wall and is saccular in type. The wisps are made of long lengths of fine dull gilt wire which can be folded up like an umbrella and put inside a small cannula. The latter is introduced into the aneurysm through a trocar which has been passed through the skin under local anæsthesia, and the wisp is pushed out of the cannula with a ramrod. Although the wire, of which it is made, is very fine the total surface area of foreign body, put into the blood in the diverticulum, amounts to several square inches. Borrie and Griffin (1950) have used this method at Mason's clinic and have claimed fairly good results. The benefit achieved was to alleviate pain and to slow up the march of the disease; but a permanent cure is not likely.

Several modifications of Colt's procedure have been tried in the management of innominate aneurysms. Mason advocated distal ligation, thus converting a fusiform into a saccular aneurysm, and then he introduced several Colt's wisps into the cul-de-sac. This approach offers promise of success but is only practicable in some arteries. Blakemore and King used insulated coin silver wire, which they put in through a special needle, and passed an electrothermic coagulating current which could be regulated at will through the wire. Each 10-meter segment was heated to 80°C. for a 10-second period and this resulted in a tenacious protein coagulum forming in the aneurysm. Blakemore reported 63 cases of syphilitic aneurysm so treated (1948) and claimed that 27 per cent were living 2–11 years after operation.

Linton relied on the amount of wire introduced and in some cases he put in as much as 150 yards into one aneurysm. He made the observation that it was of no consequence

Radiologically

An aortic aneurysm presents in ordinary films as a dense circumscribed tumour based upon the mediastinum or the heart. The mass may be roughly circular, elongated, or lobulated. It may be confined to one side of the mediastinum or may reach into both lung fields. In its walls calcification or ossification may be seen in penetrating films or tomograms. The latter are particularly important in working out the anatomy of all doubtful mediastinal shadows. The bones of the chest wall may show erosion; this is often limited to one area and, in the case of the vertebral column involves only one side of adjacent vertebral bodies.

The most important discussion has ranged about pulsation. The facts are these. When an aneurysm comes up towards the skin visible and palpable expansile pulsation occurs, and becomes more and more obvious as the end approaches. In sharp contrast to this an aneurysm generally cannot be seen to pulsate on screening and often shows no evidence of movement of kymography. on the other hand it may pulsate a little in one place and not in another. This is no more than one might anticipate from a knowledge of the pathology of the lesion; and some surgeons, working in this field, will have had the humiliating experience of trying to remove an aneurysm on the mistaken assumption that it was a mediastinal tumour. If a circumscribed tumour in the mediastinum pulsates on screening or kymography, the tumour is probably a benign cyst or a lipoma situated against the pericardium and the pulsation is directly transmitted from the heart.

Angiocardiography reveals the anatomy of difficult cases. It can not only prove that a tumour is an aneurysm but it can show the thickness of the walls and the amount of clot inside the sac. It should be a routine procedure whenever an operation is contemplated.

A thoracic aneurysm may produce changes in the lung due to progressive bronchial occlusion.

Aortography

Before surgical treatment of a thoracic aneurysm can be discussed it is essential that an aortogram be available. This is made either by passing a catheter, in a retrograde fashion, up the brachial artery so that the tip lies in the arch of the aorta and a liquid, opaque to X-rays, is then injected quickly, and several films are taken in quick succession. Alternatively, the catheter can be passed through a canula inserted percutaneously into one of the carotid arteries, and passed from there down into the aortic arch. The making of thoracic aortograms may be dangerous if the dye is injected either into the coronary arteries or passes into the cerebral vessels. The resultant X-rays show not only the type of aneurysm (e g. saccular, fusiform, etc.) but its exact anatomical site and the presence or absence of other aneurysms which may have escaped detection on the ordinary radiographs.

Bronchoscopy

This investigation will be necessary in the assessment of most cases. Sometimes an aneurysm bursts spontaneously into the air passages, and the surgeon will remember that he may precipitate the event by passing a bronchoscope. It is also relevant that pulsation can often be detected bronchoscopically when the cause is not an aneurysm; the usual reasons for this mistake are the presence of a vascular malignant tumour, or of hypertension with an unwound aorta, and sometimes the presence of a vascular ring or abnormal position of the arteries arising from the aorta (q.v.).

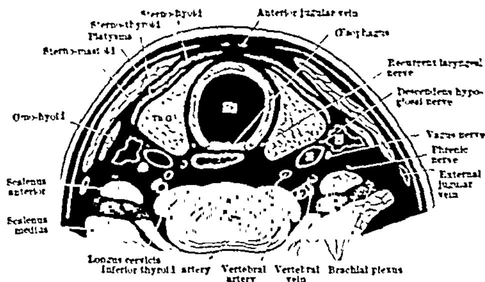
CHAPTER XII

THE ŒSOPHAGUS

R. H. FRANKLIN

ANATOMY AND PHYSIOLOGY OF THE ŒSOPHAGUS

THE œsophagus starts at the lower border of the cricoid cartilage and ends at the cardiac orifice of the stomach at the level of the eleventh thoracic vertebra. In the adult it is about 25 cm. (10 in.) long. The distance from the anterior alveolar margin to the cardiac orifice of the stomach is 40 cm. (16 in.). The œsophagus takes a slightly curved course. From its origin in the midline it passes slightly to the left at the root of the neck, back to the midline at the level of the fifth thoracic vertebra, and again to the left to reach the



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG. 325. Section through the neck at the disc between the seventh cervical and first thoracic vertebra
Tr. Trachea. Th G. Thyroid gland. J. Internal jugular vein. C. Common carotid artery

œsophageal opening in the diaphragm. It also curves slightly in an antero-posterior direction to correspond with the adjacent parts of the vertebral column. During its course the œsophagus is in contact with many important structures in the neck and thorax, and these are best appreciated by reference to the accompanying transverse sections (Figs. 325 to 331).

Particular attention is drawn to its relationship to the arch of the aorta. In this situation the œsophagus lies behind and to the right of the arch and is closely applied to the arch, the termination of the trachea and to the left bronchus. The left recurrent laryngeal nerve and branches passing to the cardiac plexus lie between the œsophagus and the aortic arch. This part of the œsophagus is also in contact with the right pleura and vena azygos

if some of it found its way into the main blood channel, because only that in the sac of the aneurysm produced clot. This may, or may not, be true.

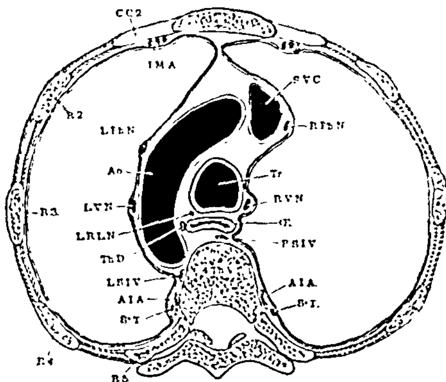
In 1943 Harrison and Chandy claimed to have cured an aneurysm of the subclavian artery by wrapping cellophane tape around the vessel. The idea of using cellophane for this purpose originated in the experiments which Page did in 1939 to produce fibrosis around the renal arteries and so to induce hypertension. This technique produced extensive perianeurysmal fibrosis (provided the correct type of cellophane was used) and was said to be especially suitable for fusiform dilatations of the thoracic or the abdominal aorta.

A recent opinion on the value of cellophane wrapping comes from Osler Abbot who reported, in 1950, that the most which could be claimed was some relief of pain in about 40 per cent of patients. He stated that the operation did not prolong life, and although it might be the only thing possible for some fusiform aneurysms, the method was not to be recommended.

The present approach to the problem of thoracic aneurysm is that advocated by Bahnson, De Bakey and others, who advise excision of the "tumour" whenever possible. Two types of cases are suitable for such treatment. The commoner is the saccular aneurysm, and here it should be possible to dissect out the neck and adjacent parts of the aorta, without touching the peripheral coverings of the aneurysm, which are thin and may rupture when dissected free from the structures they impinge upon. Having freed the neck, a suitable clamp can be placed across it, and the sac of the aneurysm cut away from the main channel of the aorta. The longitudinal incision in the latter can then be closed with sutures and, when this has been done, the sac can be excised completely. In this way very large aneurysms have been successfully removed without interrupting the flow of blood through the aorta. Excellent results have been claimed.

The second method of excision is suitable for localized, fusiform, aneurysms; and for some saccular types. The operation is begun by providing a temporary by-pass so that the blood can be diverted around the aneurysm and the pathological area can be clamped off completely from the main circulation. The by-pass can be a plastic tube anastomosed temporarily to the aorta above and below, or can be a length of human, or of pig's aorta. The aneurysm is then excised and the gap bridged by putting in a permanent arterial graft. This method has been used on many occasions with success.

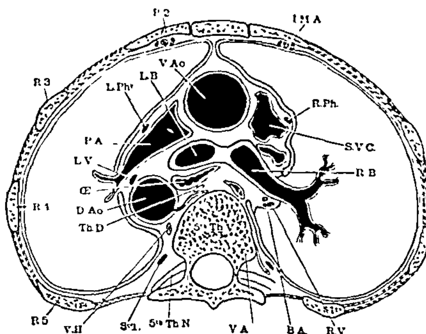
Operations of this type are exacting to the surgeon and dangerous to the patient; but a thoracic aneurysm is a most unpleasant lesion to harbour, and the relief of symptoms provided by successful operation is dramatic. Bahnson stresses the opinion that in most of the cases he has treated the aneurysm was the only dangerous lesion from which the patient suffered.



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG. 327. Section through the fourth thoracic vertebra behind and the upper part of the second costal cartilage in front

I.M A Internal mammary artery S V C. Superior vena cava. R.Ph.N Right phrenic nerve L Ph N Left phrenic nerve Ao Aortic arch. Tr. Trachea. L.V.N Left vagus nerve R V N Right vagus nerve. L.R.L.N. Left recurrent laryngeal nerve Th D Thoracic duct. Œ. Œsophagus. R S I V. Right superior intercostal vein L S I V Left superior intercostal vein. Sc T Sympathetic trunk



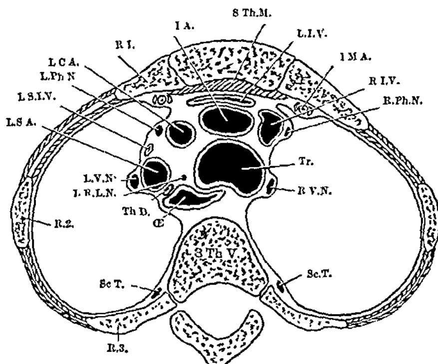
(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG 328 Section through the lower part of the second costal cartilage in front and the fifth thoracic vertebra behind

I M A Internal mammary artery V.Ao Ascending aorta. D.Ao. Descending thoracic aorta L.B. Left bronchus. R.B Right bronchus. L.Phr. Left phrenic nerve R.Ph Right phrenic nerve. S.V.C Superior vena cava. P.A. Pulmonary artery (in front of the right bronchus another branch of this is seen) L.V. Left vagus R V. Right vagus. Œ. Œsophagus. Th D. Thoracic duct. V A. Vena azygos V.H Vena hemiazygos. B.A. Bronchial artery. Sc.T. Sympathetic trunk.

arch. The vagus nerves come in contact with the œsophagus below the roots of the lungs, the right nerve lying behind and the left in front.

The œsophagus lies in a bed of loose areolar tissue which allows considerable movement in a lateral direction. The muscular coat of the œsophagus consists of an outer longitudinal and an inner circular layer, and between the two layers of muscle is connective tissue containing Auerbach's plexus and blood vessels. The mucous membrane



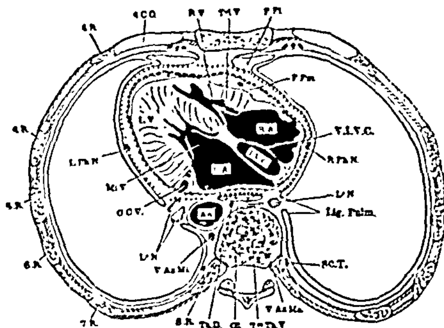
(*From the "Surgery of the Œsophagus," by R. H Franklin. Edward Arnold & Co*)

FIG 326 Section through the first rib in front and the third thoracic vertebra behind

S Th M Sternothyroid origin L I V. Left innominate vein. I M.A. Internal mammary artery. R.I V Right innominate vein R.Ph N. Right phrenic nerve L Ph N Left phrenic nerve. I A Innominate artery. L C A. Left carotid artery L S A Left subclavian artery. L.S.I.V. Left superior intercostal vein L.V N Left vagus nerve R.V N Right vagus nerve. L R L N Left recurrent laryngeal nerve Tr Trachea. Œ Œsophagus. Th D Thoracic duct Sc T. Sympathetic trunk

of the œsophagus is pink-coloured at the upper end and becomes paler lower down, and at the cardia the nearly white œsophageal epithelium gives place abruptly to the red uneven-looking gastric mucosa. The mucous membrane consists of stratified squamous epithelium, connective tissue and the muscularis mucosæ. In the region of the cardia are a number of glands which may enlarge after middle age and produce irregular masses of gland tissue Between the mucous membrane and the muscle lies a submucous layer which is continuous with that of the stomach. This layer contains small compound racemose glands each of which opens on to the surface by a long duct Meissner's plexus lies in this layer which also contains numerous lymphatic channels.

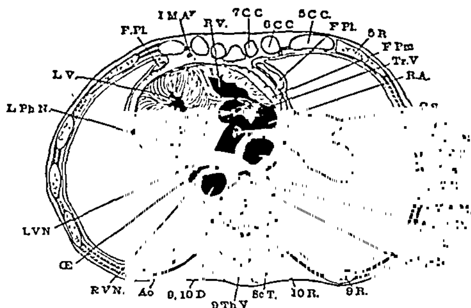
Blood Supply. The blood supply of the œsophagus comes from the inferior thyroid, subclavian, bronchial, left gastric and inferior phrenic arteries and from the aorta. The œsophageal veins drain upwards into the systemic system and at the lower end into the portal system.



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG 330 Transverse section through the thorax at the level of the fourth costal cartilage

R.V Right ventricle Tri V. Tricuspid valve F Pl Fatty fold in the pleura. F Pm. Fat deep to the visceral layer of the pericardium R.A Right atrium. L A Left atrium. V I V C Valve of inferior vena cava. I V C. Inferior vena cava R Ph N Right phrenic nerve L Ph N Left phrenic nerve. Mi.V. Mitral valve G C V. Great cardiac vein. Lc.N. Lymph nodes. Ao. Descending aorta V.H Vena hemiazygos Th D Thoracic duct. Œ. Œsophagus V Az Vena azygos Sc T. Sympathetic trunk. Lig Pulm. Ligamentum pulmonis

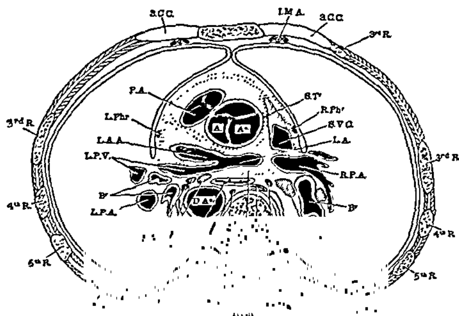


(From the "Surgery of the Œsophagus," by R H Franklin, Edward Arnold & Co.)

FIG 331 Section through the disc between the ninth and tenth thoracic vertebræ behind and xiphisternal junction in front.

I M Ay Internal mammary artery F Pl. Fatty fold in pleura. F.Pm. Fat in the visceral pericardium Tr.V. Inferior cusp of the tricuspid valve. R.A. Right atrium C S Coronary valve R Ph N Right phrenic nerve. L.Ph N. Left phrenic nerve R V Right ventricle L V. Left ventricle. R V.N. Right vagus nerve L V N Left vagus nerve I V C Inferior vena cava Dia. Right cupola of diaphragm exposed V A. Vena azygos Thc.D Thoracic duct. Œ Œsophagus. Ao Descending thoracic aorta. G Spl.N Greater splanchnic nerve. Sc T Sympathetic trunk. The parietal pericardium is represented by a dotted line.

Nerve Supply. The nerve supply is derived from the sympathetic and para-sympathetic systems, the fibres of the latter being carried in the vagi. The œsophagus is surrounded by the plexus gulæ and in the superior mediastinum this is formed by branches of the right vagus and left recurrent laryngeal nerves, and branches from the upper thoracic sympathetic ganglia. In the posterior mediastinum, branches from both vagi and from the



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG. 329. Transverse section through the thorax at the level of the third costal cartilage

I M A Internal mammary artery. S Tr. Sinus pericardii transversus. P A Pulmonary artery and valve. A.Ao. Ascending aorta and valve R Ph Right phrenic nerve. L.Ph. Left phrenic nerve. S V C. Superior vena cava opening into the right atrium. L.A. Left atrium. L.A.A. Left auricular appendage. L P.V. Left pulmonary vein. R.P.A. Right pulmonary artery. L.P.A. Left pulmonary artery. Br. Bronchus Br.N. Bronchial lymph nodes D.Ao. Descending aorta Œ. Œsophagus Sc T. Sympathetic trunk. Th.D. Thoracic duct S.Ob. Sinus obliquus. R.V. Right vagus. L.V. Left vagus. V.Az. Vena azygos.

splanchnic nerves make up the plexus. Division of the vagi produces a temporary spasm of the cardia.

Lymphatic drainage. The submucous layer is freely supplied with lymphatic vessels which pass from the muscle of the œsophagus and run upwards and downwards in the adventitia, where there are a few lymph nodes. Drainage then takes place to the lymph nodes of the posterior mediastinum and to the nodes on the lesser curvature of the stomach and around the cœliac axis.

Sphincters of the Œsophagus. The cricopharyngeus muscle forms the upper sphincter and the purpose of this is to keep the gullet closed except during the swallowing of food or liquids and so prevent it filling up with air on inspiration. The lower or cardiac sphincter serves to prevent constant regurgitation of gastric contents into the œsophagus. The exact way in which this sphincter works is not quite clear, but the nature of the insertion of the œsophagus into the stomach is one important factor in producing the sphincteric effect. At the cardiac orifice the right border of the œsophagus passes on to the lesser curvature of the stomach without any appreciable change in direction, whereas

type of abnormality accounts for 80 per cent of the cases seen. In most of the remainder there is a blind upper and lower œsophageal segment. Very exceptionally there may be a fistula between the upper segment and the trachea, or between both segments and the trachea, and on very rare occasions a tracheo-œsophageal fistula without atresia (Fig. 333). Congenital narrowing of the œsophagus has been described and also complete absence of the œsophagus, but all these abnormalities are comparatively unimportant compared with atresia with or without tracheo-œsophageal fistula. The so-called congenital short œsophagus is often a misnomer and will be considered later.

CONGENITAL ATRESIA OF THE ŒSOPHAGUS

This condition was recognized and described in 1697 by Gibson, who was an army physician and a grandson of Oliver Cromwell. From time to time since that date the condition has been referred to. In 1910 Sir Arthur Keith collected 14 examples of the condition in the museums of London and made an accurate description of the anomaly.

Sporadic attempts at surgical treatment were made but most of these consisted in carrying out a gastrostomy. This procedure was doomed to failure because the communication between the lower œsophageal segment and trachea inevitably led to the respiratory system being flooded with gastric contents, and even in the less usual type of anomaly in which no fistula exists between the œsophagus and trachea, the blind upper segment of œsophagus fills up with mucus which spills over into the lungs and produces an aspiration pneumonia. The fact that the lung is so readily affected in these cases has led to a diagnosis of atelectasis, pneumonia, or some form of cerebral injury, and the infant has died without the true diagnosis being considered. The supposed rarity of the condition has been another factor in preventing the development of surgical treatment; in fact, the condition probably occurs about once in every 2,500 births.

A recognition of the surgical problems involved led to the evolution of treatment by multiple operations, and a number of ingenious attempts were made to overcome the difficulties One of these was to bring the stomach out on to the surface of the abdomen rather in the way in which the colon is brought out for a colostomy. The stomach was opened or cut across and the proximal end used to drain the fistula and the lower end to feed the child. At a later date further operations were carried out to construct an antethoracic œsophagus. The first successful operations of this nature were completed by Ladd in 1939 and Leven in 1940. It was soon realized that primary anastomosis, if it could be achieved, was the most satisfactory method of dealing with the problem and an attempt had been made by Lanman in 1936, but without success. In 1941 Haight and Towsley reported a completely successful direct anastomosis carried out by the extra-pleural route, and Haight's early results were repeated both by himself and other surgeons.

Advances in anæsthesia and the introduction of antibiotics have enabled the trans-pleural route to be used with greater safety than was previously possible and with less difficulty than is experienced when the extra-pleural approach is made. The details of the operation will be discussed later.

Diagnosis. It is most important that the possibility of this abnormality should be well known to midwives and pædiatricians who have the first care of small infants. Excessive mucus in the nasopharynx is commonly noted at birth and may give rise to cyanosis, but the cyanosis is improved by a single aspiration of the nasopharynx and the condition does

the left border forms an acute angle with the stomach which can be still further diminished by the action of the oblique muscle fibres of the stomach. This mechanism results in the closure of the lower end of the œsophagus when the fundal end of the stomach undergoes distension. Other factors concerned are the muscle fibres of the right crus producing the so-called "pinch-cock" action, and the mucous membrane of the lower end of the œsophagus which may form a valve comparable with that produced by a "glove-finger" drain.

Movements of the Œsophagus. The passage of food down the œsophagus takes place chiefly by peristaltic movements and to a certain extent by gravity.

(1) PRIMARY WAVE. This is a true peristaltic wave which flows from the upper end of the gullet right down to the cardia, being preceded by a wave of relaxation.

(2) SECONDARY WAVE. This starts at the level of the aortic arch and serves to reinforce the primary wave.

(3) TERTIARY CONTRACTIONS. These may occur in the lower two-thirds of the œsophagus and may last for several seconds. They do not appear to serve any useful function and may lead to mistakes in diagnosis.

The bolus of food usually takes about 5 seconds to reach the lower end of the œsophagus.

DEVELOPMENT

The œsophagus develops in the primitive foregut. Two lateral grooves which run longitudinally, meet internally and join to separate the trachea and larynx in front from



(From the "Surgery of the Œsophagus," by R H Franklin, Edward Arnold & Co)

FIG 332 Atresia of the œsophagus with tracheo-œsophageal fistula The most common type (Type III (b) according to Vogt's classification); 80 per cent of all cases fall into this group



(From the "Surgery of the Œsophagus," by R.H Franklin, Edward Arnold & Co)

FIG 333. A rare anomaly. Fistula between œsophagus and trachea without atresia.

the œsophagus behind. The fusion of the two lateral ridges should be completed by the sixth week of intra-uterine life. Failure of these ridges to join, or an oblique arrangement of the ridges, will explain the deformities encountered. The most common abnormality is atresia of the œsophagus with a tracheo-œsophageal fistula (Fig. 332). This

Operation by the Right Trans-pleural Approach

This is the standard approach for the common type of anomaly in which there is a blind upper segment and in which the lower œsophageal segment arises from the trachea close to the bifurcation The operation is carried out under general anæsthesia administered through an endotracheal tube The introduction of an endotracheal tube is most



(From the "Surgery of the Œsophagus" by R H Franklin Edward Arnold & Co.)

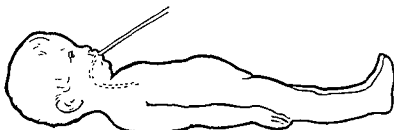
FIG 335 The introduction of 0 5 c c of iodised oil confirms the presence of a blind upper segment

important because it enables the anæsthetist to aspirate the bronchial tree and makes it possible to leave the lungs fully expanded at the end of the operation.

An intravenous drip is set up before starting the operation. The infant is laid on the left side and an assistant holds the right arm so that the scapula may be drawn forward as required and the position altered if necessary. The skin incision follows the vertical border of the right scapula and is curved forwards to the mid-axillary line. The incision is deepened to divide the muscle and the scapula is retracted forwards. An intercostal

not recur. If a recurrence of cyanosis takes place shortly after aspiration, the possibility of atresia of the œsophagus should be considered. If the condition is untreated and attempts are made to feed the child, the symptoms are accentuated; the child sucks strongly but is overcome by spluttering and cyanosis, and regurgitation of the feed. Each attack of this nature results in part of the feed entering the trachea and lungs and diminishes the chances of successful operation, and for this reason the diagnosis must be suspected at the earliest possible moment.

Any suggestion of an œsophageal abnormality should lead to the passage of a well-lubricated rubber catheter through the mouth and down the œsophagus. If the catheter is constantly arrested at a distance of 10–12 cm. from the anterior alveolar margin, the diagnosis of œsophageal atresia is almost certain (Fig 334). Great care must be taken not to use too fine a catheter in carrying out this test, or it may curl back and give rise to the impression that the stomach is being entered.



(From the "Surgery of the Œsophagus," by R. H Franklin, Edward Arnold & Co)

FIG 334 The first step in the diagnosis of œsophageal atresia A well-lubricated rubber catheter is introduced through the mouth and its arrest 10–12 cm. from the alveolar margin is almost diagnostic of the condition.

Confirmation of the Diagnosis. The best plan is to leave the confirmation of the diagnosis until the child can be transferred to a centre which is properly equipped for both the radiological diagnosis and the operative treatment of the condition The few hours' delay which this may involve is of no serious consequence to the outcome, whereas hasty attempts at radiological confirmation may spoil the chances of successful surgery. In the hours which may intervene between the suspicion of the diagnosis and the infant's admission to such a centre, care must be taken to keep the blind upper segment empty by aspiration and to change the infant's position frequently so that chest complications are kept to a minimum. Antibiotics should be administered by injection.

The diagnosis is confirmed radiologically and if this examination is carried out accurately, the type of deformity present can also be ascertained. The infant is screened and the state of the lungs carefully noted, and the presence of air in the stomach and intestines looked for. If air is present in a case of atresia, it means that there must be a communication between the lower segment and the trachea After the preliminary inspection, a rubber catheter is introduced through the mouth to the lower end of the blind upper segment, and ½ ml. of iodised oil is introduced. If atresia is present, the appearance of [illegible] segment is characteristic (F[illegible] 335). The iodised oil is watched for a short [illegible] segment and the trachea, and [illegible] ination both the presence of atresia and the type of the deformity can be diagnosed.

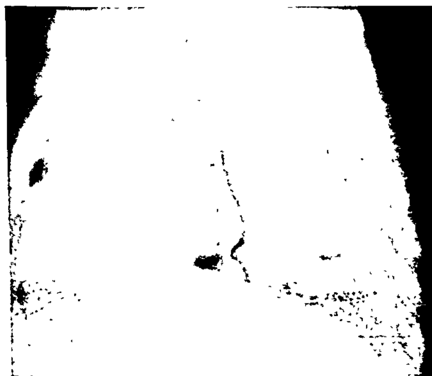
in the anastomosis occurs, it may be necessary to retain the intercostal drainage for a longer period, or even to reintroduce it. If the leak persists, a gastrostomy should be made before the child's condition deteriorates

Operations for Infants Suffering from the Type II Deformity

In this type of deformity both the upper and lower segments end blindly and very often the lower segment is so short that there is no chance of making an anastomosis If the right-sided approach has been used, nothing can be done except to carry out the first stage of a multiple-stage operation To avoid this it may be justifiable if this type of deformity has been diagnosed, to make a left-sided approach so that in the event of the lower segment being very short or absent, the stomach can be used to complete the anastomosis.

CONGENITAL SHORT ŒSOPHAGUS AND REFLUX ŒSOPHAGITIS

A condition is found in both infants and adults in which the œsophagus appears to stop above the diaphragm where it joins the intra-thoracic portion of stomach (Fig 336) It is remarkable, however, that although the condition is seen in infants and small



(From the "Surgery of the Œsophagus" by R H Franklin, Edward Arnold & Co.)

Fig 336 "Short œsophagus" in an infant of eighteen months

children and again in middle-aged patients, it is much more rarely seen in late childhood and early adult life, and for this reason it seems probable that many cases of so-called congenital short œsophagus are really examples of an acquired condition.

The most likely cause of a short œsophagus appears to be spasm of the longitudinal muscle of the gullet due to œsophagitis, and this in turn is the result of a weak œsophageal sphincter, allowing gastric contents to regurgitate. The pathological process set up in

incision is made in the fourth interspace and a self-retaining retractor introduced. The lung is inspected and if there are collapsed areas, it is a good plan at this stage to massage the air gently into these parts so that full expansion is obtained. If a considerable part of the lung is collapsed, it may indicate either that there is an accumulation of mucus in the bronchial tree or that the endotracheal tube is not correctly in place. These points must be attended to. The lung is now allowed to collapse and the first landmark identified, which is the vena azygos arch. This represents the presumed level of both the lower end of the blind upper segment and the point of communication of the lower segment with the trachea. It is usually necessary to ligature and divide the azygos arch in order to obtain a clear view. In ligaturing this vein, thread ligatures are used introduced on an aneurysm needle, and sufficient space must be left between the two ligatures so that there is no risk of one coming off when the intervening portion of vein is divided.

The blind upper segment can usually be identified easily, but if there is any difficulty a rubber catheter passed into the segment from the mouth will demonstrate its position. Stay sutures are then introduced so as to manipulate it without any unnecessary handling. The point where the œsophagus arises from the trachea may be more difficult to determine. When this point has been identified the fistulous communication is divided and the part opening into the trachea is closed with a fine silk stitch. This stitch should be of 5/0 silk carried on a very small, round-bodied, curved needle. Before dividing the fistula the lower segment must be controlled with stay sutures or there may be a tendency for it to retract downwards. An opening is made in the blind upper segment and interrupted sutures, using the same suture material as before, are introduced through all coats of both segments. It should be the aim to pass 4 interrupted sutures in this way before any of them are tied. On tying these the posterior part of the two segments is brought into apposition. A fine rubber catheter is passed down into the stomach and up through the upper segment into the mouth where it is recovered. The introduction of this catheter makes it a comparatively easy matter to introduce another 3 or 4 interrupted sutures through all coats until the anastomosis is completed. The stomach should be aspirated through this catheter and very often a considerable amount of air is withdrawn in this way. The catheter is then carefully pulled out through the mouth. An intercostal drainage tube is placed in position and connected with a water-seal drainage bottle, the lung is fully expanded and the chest closed. It may be advisable to give a blood transfusion towards the end of the operation. It is usually not necessary to give more than 60–80 ml.

Post-operative Care

The child should be nursed in an oxygen box for the first few hours, but as soon as breathing appears to be maintained easily the box should be removed. A careful watch must be kept on the infant because choking with mucus can occur post-operatively. Aspirations must be carried out in the same way as was done pre-operatively, but with the difference that in order to avoid damage to the anastomosis the catheter used for aspiration should have a mark 8 cm. from the end, and this mark should not be allowed to pass beyond the anterior alveolar margin The position of the infant should be changed every hour and antibiotics administered. Hydration is maintained intravenously for the first 48 hours and after this time cautious mouth feeding is started.

X-ray examinations are made of the chest post-operatively and if the lung is completely expanded, the intercostal tube should be removed on the fourth or fifth day. If a leak

or lies down, gastric contents will flow into the œsophagus and if these gastric contents are irritating, œsophagitis is set up. Many people have a hiatus hernia without developing œsophagitis.

The pathological change associated with œsophagitis is a superficial ulceration which affects principally the lower part of the œsophagus, but which may gradually involve a large part of the gullet. The ulceration shows a tendency to heal and break down.

The course of the disease may be affected by treatment and sometimes the condition resolves spontaneously but in general, if no treatment is given, the condition tends to become worse. As the result of the ulceration the patient suffers from a steady loss of blood and the associated spasm causes shortening of the œsophagus by contraction of the longitudinal fibres and dysphagia due to contraction of the circular fibres. The result of this is that more and more of the stomach becomes drawn up into the chest and the sphincteric mechanism is still further weakened. The shortening of the œsophagus and the stricture formation are at first entirely due to spasm, but later on fibrous changes occur.

Symptoms. The principal symptoms of reflux œsophagitis are pain, dysphagia, and symptoms caused by anæmia. All these symptoms may occur at the same time, or one or other may be absent.

The pain is characteristic and consists of heartburn, which is felt behind the sternum after eating or drinking, and the pain may radiate up into the neck and jaw and down one or both arms, and sometimes through to the back. Characteristically the pain is made worse by lying down, bending or, in fact, carrying out any movement which tends to favour the reflux of gastric contents into the œsophagus.

The dysphagia which is produced is usually subjective in the first instance, but if the condition progresses and a permanent stricture forms, regurgitation of food eventually occurs.

Anæmia results from the gradual blood loss over a long period and unless the stools are examined for occult blood there may be no indication that hæmorrhage is occurring in the alimentary tract. Severe bleeding resulting in hæmatemesis or melæna is occasionally seen, but its occurrence in these patients is usually an indication that a chronic gastric ulcer has formed in the thoracic stomach (Fig. 338). In infants the condition may present itself in such a way that an erroneous diagnosis of pyloric obstruction is made.

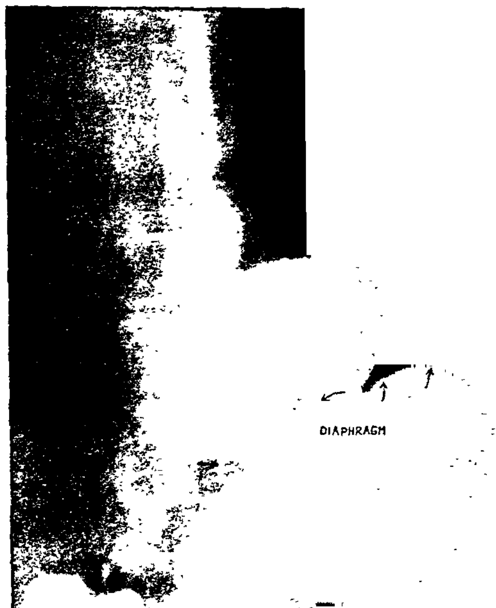
Diagnosis. The history is most important and if pain is a feature it will often indicate the correct diagnosis. Dysphagia may suggest a malignant cause in the older age group although the length of history and the occurrence of pain may favour simple ulceration, but there is no single fact in the history which can establish the diagnosis firmly and it is essential to proceed to radiological and œsophagoscopic examination.

X-ray examination may show the presence of a hiatal hernia, but this may only be seen in the Trendelenburg position and it is possible for a patient to suffer from reflux œsophagitis as the result of a weak sphincteric mechanism without any herniation being demonstrable. In some instances the radiological appearance may closely simulate a carcinoma and in others may resemble a true cardiospasm.

Œsophagoscopic Appearance

In a typical example of reflux œsophagitis the lower part of the œsophagus is reddened and may show superficial ulceration. The ulcerated area may extend a considerable

these circumstances has been appropriately named by Barrett reflux œsophagitis. Excessive regurgitation of gastric juice may occur in the vomiting of pregnancy and in connection with certain cerebral conditions, but the most common cause is incompetence



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG. 337. Hiatal hernia in a woman of sixty

of the lower œsophageal sphincter. The mode of action of this sphincter has been considered earlier.

Any condition which tends to weaken the lower œsophageal sphincter will, in the presence of increased abdominal pressure, lead to a gastric reflux. The most common finding in these cases is the presence of a hiatal hernia in which the cardia is displaced upwards and the angle of insertion of the œsophagus into the stomach is altered so that a cone of stomach projects into the chest and is joined at its summit by the gullet (Fig. 337). Under these circumstances there is no effective sphincteric action and if the patient stoops

really reflux œsophagitis, conversely a carcinoma of the œsophagus or stomach may be masked by an overlying spasm of the œsophagus

Treatment. Conservative treatment should be given a trial in all cases unless the degree and extent of the stricture formation make it clear that operation will have to be undertaken. The most important part of the conservative treatment is to impress on the patient the necessity for avoiding all positions which favour reflux from the stomach. The head and shoulders should be supported on 3 or 4 pillows at night or the head of the bed may be raised on blocks. Bending and stooping movements are to be avoided. The diet should be simple and meals taken at frequent intervals so as to avoid the risk of an empty stomach with a high acid content. Alkali is best administered by sucking alkaline tablets between meals and, in addition, olive oil should be taken at night.

In the case of infants it is usually necessary to employ either a harness or some form of box so that the child may be kept in a sitting position.

Feeding by means of a Ryle's tube is sometimes called for in difficult cases in both infants and adults.

Under this regime 70-80 per cent of the patients will improve to such an extent that no operative treatment is necessary.

If a small stricture is present, it is sometimes possible to treat this by dilatation with a gum-elastic bougie and at the same time carry out the regime described above. If dilatation of a stricture is carried out in this way, it is most important to insist on the patient taking alkalis and olive oil or there is a risk that further œsophagitis will follow at a higher level.

Operative Treatment

This is indicated when medical treatment has failed, if there is more than a slight degree of stricture formation or when there is a duodenal or gastric ulcer present. The type of operation to be carried out depends upon the pathological conditions present.

(a) Operation to Limit Regurgitation and to Diminish the Gastric Acidity

This is indicated when there is no gross stricture formation and no associated peptic ulcer. The operation is carried out as follows: A left-sided abdomino-thoracic approach is made, resecting the eighth rib. The left pulmonary ligament is divided and this discloses a cone of stomach ascending into the chest. The diaphragm is incised in a radial manner keeping well away from the right crus. A hand is introduced through the opening in the diaphragm, the size of the hernia assessed and the level of attachment of the phrenico-œsophageal ligament demonstrated. This attachment is taken as indicating the junction between the œsophagus and cardia, and one of the aims of the operation is to attach it to the undersurface of the diaphragm so as to keep the whole of the stomach in the abdomen. The œsophagus is now mobilized, taking care not to destroy the phrenico-œsophageal ligament. To help in the mobilization and to reduce the acidity both vagi are divided. The pyloric sphincter is divided, taking care not to open the mucous membrane. The stomach is now kept in the abdomen by suturing the phrenico-œsophageal ligament to the diaphragm in several places. The incision in the diaphragm is closed and no attempt is made to narrow the hiatus which will rapidly adapt itself to the size of the œsophagus.

distance upwards, but it is always worse at the lower end. If dysphagia has been present, there may be some fluid in the œsophagus and when this is removed by suction there is a tendency for the gullet to fill up again from the stomach. If shortening of the œsophagus has occurred, it will be evident by measuring the distance from the teeth to the œsophago-gastric junction, but a difficulty may arise here in that the ulcerated œsophagus may make



(From the "Surgery of the Œsophagus," by R H Franklin, Edward Arnold & Co)

FIG 338 Hiatal hernia with a chronic gastric ulcer in the thoracic portion of the stomach.

it hard to determine the precise line of junction with the stomach. In the case of true cardiospasm the œsophagus may contain large quantities of food or fluid, but when this has been removed there is no tendency for it to accumulate again during the examination. Another point of difference between these two conditions is that ulceration is rare in true cardiospasm, and if it does occur it is comparable with the stercoral ulcers found in the colon and may occur at any level.

Just as X-ray examination may suggest a carcinoma when in fact the condition is

really reflux œsophagitis, conversely a carcinoma of the œsophagus or stomach may be masked by an overlying spasm of the œsophagus.

Treatment. Conservative treatment should be given a trial in all cases unless the degree and extent of the stricture formation make it clear that operation will have to be undertaken. The most important part of the conservative treatment is to impress on the patient the necessity for avoiding all positions which favour reflux from the stomach. The head and shoulders should be supported on 3 or 4 pillows at night or the head of the bed may be raised on blocks. Bending and stooping movements are to be avoided. The diet should be simple and meals taken at frequent intervals so as to avoid the risk of an empty stomach with a high acid content. Alkali is best administered by sucking alkaline tablets between meals and, in addition, olive oil should be taken at night.

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(a) Operation to Limit Regurgitation and to Diminish the Gastric Acidity

This is indicated [illegible]

ulcer. [illegible]

is made [illegible]

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(b) Partial Gastrectomy and Replacement of the Stump of the Stomach below the Diaphragm

This procedure is suitable in those patients who have symptoms of reflux œsophagitis associated with a duodenal or gastric ulcer which is not within 2 in. of the cardia. The surgical approach, the mobilization of the œsophagus above the phrenico-œsophageal ligament and the division of the vagi are carried out as described above. The stomach and duodenum are examined carefully, having made a radial incision in the diaphragm. A partial gastrectomy is carried out suitable to the position and extent of the ulcer found, i.e. if the ulcer is duodenal, a Polya type of operation is usually chosen, if a gastric ulcer is present, a Billroth I gastrectomy is employed. The stump of the stomach is now kept in place below the diaphragm by suturing the phrenico-œsophageal ligament as described before.

(c) Partial Œsophagectomy together with Total Gastrectomy. Restoration of Continuity by Roux en Y Anastomosis

This procedure is suitable in those cases in which there is a severe stricture of the lower end of the œsophagus which does not extend higher than a point midway between the diaphragm and the arch of the aorta. The presence of a gastric or duodenal ulcer, together with severe symptoms of reflux œsophagitis, is also an indication. A left-sided abdomino-thoracic approach is made as already described. It is an advantage to mobilize the small bowel for the Roux en Y before carrying out the resection of the stomach and lower œsophagus. In this way it is possible to be sure that the mobilized segment of small bowel is viable. The mobilization is carried out as follows: Clamps are applied to the small bowel at a point about 8 in. from the duodeno-jejunal junction and the bowel held out on either side of the clamps by an assistant. The exact position of the clamps is adjusted so that an incision can later be made between the blood vessels leading down to the vascular arch. The bowel is not divided at this stage. By having a white mop behind the mesentery or, if necessary, by using a light, it is possible to see the position of the blood vessels. The peritoneum of the mesentery on the side nearer to the operator is carefully divided parallel to the vascular arch thereby exposing the tributaries and branches. These are now carefully picked up with an aneurysm needle and tied with fine thread and divided, the arch itself being divided at the selected point opposite to the clamps. The peritoneum of the mesentery on the surface further from the operator is then divided and the bowel itself is cut across between the clamps. An estimate is made to see if the mobilized segment of bowel will reach as high as the proposed line of section of the œsophagus. If it appears to be satisfactory the ends of the mobilized intestine are carefully wrapped in moist packs and put aside until they are required. Attention is now directed to resecting the stomach and lower œsophagus. The short gastric vessels are divided and the division is extended all the way along the gastro-colic omentum until the duodenum is reached. The stomach is held over to the patient's right side and the left gastric vessels are exposed, ligatured and divided. The right gastric vessels are similarly divided and clamps are then applied to the duodenum. The duodenum is divided and the duodenal stump closed and invaginated. The stomach is now turned upwards into the chest and the remains of the lesser omentum are secured in forceps and ligatured and divided as are any vessels entering the surface of the œsophagus from the

undersurface of the diaphragm. The lower part of the œsophagus is separated from the surrounding pleura and to increase the mobility both vagi are divided. Stay sutures are placed in the œsophagus at either side of the proposed line of section. A clamp is not used on the œsophagus. The actual division of the œsophagus must be made carefully in order to keep the mucous membrane within easy reach To achieve this the muscle is first divided, taking care not to cut into the lumen. By drawing gently on the stomach a band of mucous membrane is exposed and the mucous membrane is then cut about ½ in. below the cut edge of the muscle. The efferent loop of small bowel is brought through an opening in the transverse mesocolon close to the ligament of Treitze and an end-to-end anastomosis carried out with the œsophagus. A satisfactory way of making the anastomosis is as follows: three interrupted mattress sutures of thread are passed through all coats of both small bowel and œsophagus, starting and finishing on the inside of the small bowel. When all three sutures have been introduced they are tied, taking care not to tie them too tightly. A continuous through and through suture of fine catgut is started on the posterior surface and passing through all coats, is carried right round to the front As it turns the corner the stitch is applied from within out on each side until it reaches its starting point. It is often impossible to introduce Lembert sutures because of the tendency to cut out from the œsophageal muscle, but in cases of reflux œsophagitis the neighbouring œsophagus is often thickened and may be easier material to suture than normal œsophagus. A trial is therefore made and if a Lembert suture seems to hold well, a series is put in all the way round the anastomosis If there is a tendency for the sutures to cut out, reliance is placed on drawing the small bowel up to the œsophagus by means of a stitch passing through the pleura The anastomosis is then further reinforced by suturing the pleura over the anastomosis to the small bowel and if necessary turning over a mobilized flap of pleura The afferent loop of small bowel is anastomosed to the side of the efferent loop. This anastomosis is placed so as to lie just below the opening in the mesocolon to which both parts of the small bowel are attached The small bowel is also attached carefully to the margins of the diaphragm and the diaphragm itself is closed, using interrupted sutures of thread and a continuous suture of catgut. A water-seal drain is introduced and the incision closed

(d) Œsophageal Resection with Restoration of Continuity by means of Œsophago-gastric Anastomosis above the Arch of the Aorta

This procedure is indicated in those cases in which there is a very extensive stricture of the œsophagus possibly reaching as high as the arch of the aorta, but associated with a healthy stomach and duodenum The advantage of using the stomach in these circumstances is that it is considerably easier and more satisfactory than the use of a very long Roux en Y mobilized segment of small bowel. Fear has been expressed in some quarters as to the wisdom of this procedure because of the danger of producing œsophagitis at a higher level. It should be noted that a small amount of stomach in the chest is more dangerous in this respect than a large portion. This is because in cases in which the œsophago-gastric anastomosis lies 1 or 2 in. above the diaphragm, respiratory movements will result in the gastric contents being drawn up into the œsophagus with possible ill effects. If, however, the œsophago-gastric anastomosis lies above the arch of the aorta so that the whole of the stomach virtually lies within the chest, respiratory movements will result in the gastric contents moving up and down but still within the stomach. It is

necessary to warn the patient to avoid postures which favour regurgitation, but provided this is done there seems to be no increased incidence of reflux œsophagitis when this procedure has been used.

The left-sided abdomino-thoracic approach is made and the stomach mobilized as has been described. The scapula is now drawn forward and an intercostal incision made in about the fourth space. This second incision facilitates the mobilization of the middle of the œsophagus and makes it possible to complete the anastomosis with safety. The œsophagus is mobilized right up to the arch and the pleura above the arch is then divided over the œsophagus and this part of the œsophagus also mobilized. The œsophagus is controlled with stay sutures as before and divided above the arch and below at the œsophago-gastric junction, and the line of section closed and carefully invaginated. An opening is made in the fundus of the stomach corresponding in size with the œsophagus and an anastomosis carried out in front of the arch of the aorta, using the same method of *applying the sutures as has been described above. The diaphragm is closed in the same* way and care taken to attach the stomach to the margins of the diaphragm. In some cases it is possible to preserve the right crus of the diaphragm during this operation and when this can be done it makes the subsequent closure easier. The pyloric sphincter is divided in order to guard against any subsequent gastrostasis and the wound is closed with a water-seal drain in the chest as has been described.

CARDIOSPASM

The term cardiospasm or achalasia should be restricted to a condition which affects the lower two-thirds of the œsophagus or that part of it which is provided with unstriated muscle. The term should not be used in those cases which show a localized spasm, the result of some irritating lesion in the neighbourhood. The exact ætiology of true cardiospasm is not properly understood, but it seems to depend upon an inco-ordination between the sympathetic and the para-sympathetic innervation or on a failure of conduction of peristalsis down the œsophagus. It is known that anything which produces over-action of the sympathetic, such as fear, may make the symptoms of cardiospasm worse, and similarly interference with the vagal supply will in some patients produce a temporary condition which is very similar to cardiospasm.

The symptoms of cardiospasm are at first subjective; the patient feels that his food sticks from time to time and then passes on. Later, actual regurgitation occurs at frequent intervals and the patient is unable to finish a meal without returning some of what he has taken. In the early stages the œsophagus is not dilated but at a later date it may become both dilated and elongated so that it takes a sigmoid shape. When gross dilatation of the œsophagus has taken place regurgitation becomes less frequent but the quantity returned is correspondingly greater. At this stage other effects are likely to follow. During sleep regurgitation of food into the respiratory passages may cause choking or coughing and ultimately may set up pulmonary changes. In a longstanding case pulmonary osteo-arthropathy may occur and the health undermined by long-continued septic absorption. Considerable dilatation of the œsophagus may be present before any symptoms referable to swallowing are noticed, and symptoms may be absent until the condition has been discovered incidentally. Men are more frequently affected than women and the condition is most often seen in the third and fourth decades, but it may occur at any age.

Carcinoma sometimes supervenes in late cases at any level of the œsophagus.

Diagnosis. On radiological examination the œsophagus is usually dilated, but the degree of dilatation shows great variation. The lower end of the œsophagus shows a smooth obstruction at the level of the diaphragm. There is no irregularity or eccentricity



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG. 339. Dilatation of the œsophagus in long-standing cardiospasm.

such as is usually seen in the case of a carcinoma. The gastric air bubble is usually absent in cardiospasm (Fig. 339).

Confirmation of the Diagnosis. Unless œsophagoscopy is contra-indicated because of extreme age or the presence of an aneurysm, this method of confirmation should always be carried out. The differences between the œsophagoscopic appearance found in this condition and reflux œsophagitis have already been described. It is particularly important not to miss an underlying carcinoma.

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Treatment. The first line of treatment is to carry out thorough dilatation by means of a Plummer or Negus hydrostatic bag (Fig. 340). This can be done conveniently at the time of the œsophagoscopic examination. To make sure that the dilatation is effective

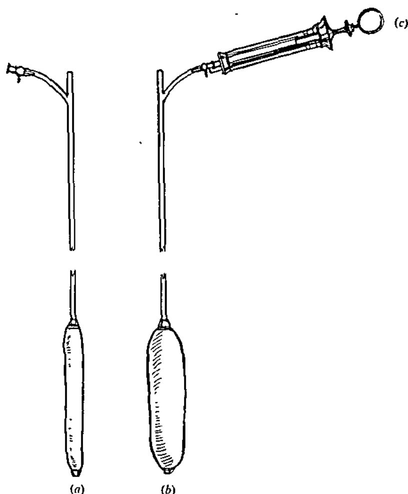


FIG 340 Negus bag
(a) Empty.
(b) Distended with water
(c) Syringe for filling the bag

it is a good plan to introduce the bag in the first instance to a point a little beyond the cardia, fill the bag with water, then empty it and repeat the process at two further points each about ½ in. higher than the other. Dilatation carried out in this way will produce a striking symptomatic improvement in a considerable number of patients. In some cases no improvement is produced and in others the horizontal course taken by the lower part of the œsophagus may make the introduction of the bag difficult.

If dilatation with the Plummer bag has proved unsuccessful or impossible to carry out, operative treatment may be required. Heller's operation carried out through a left thoracotomy incision is a very satisfactory procedure. The aim is to divide the muscles down to the mucosa over the distal 1½ in. of the gullet and proximal inch of the stomach. To do this it is necessary to draw a small cone of stomach up into the chest, but care should be taken to disturb the hiatus as little as possible. At the end of the operation the œsophagus is sutured to the right crus of the diaphragm so that the whole of the stomach

lies in the abdominal cavity. To make a complete division of the muscle without damaging the mucous membrane may be difficult, but if the operation is carried out meticulously the results are excellent. The abdominal approach is sometimes more difficult and there is a greater risk of producing a hiatal hernia or of carrying out an inadequate division of the muscle.

Elderly patients are encountered in whom it seems unwise to carry out an œsophagoscopic examination, and in these there is still a place for the use of Hurst's mercury bougies The disadvantage of this method is that the mercury bougie has to be passed at least every day and although the average patient finds little difficulty or inconvenience in doing so, this constant repetition makes the method undesirable save in exceptional circumstances

PHARYNGEAL DIVERTICULUM

A pharyngeal diverticulum arises as a herniation in the midline of the pharynx posteriorly at a point on the pharyngeal wall which is unsupported by muscle, lying between the oblique constrictor fibres and the circular muscles constituting the crico-pharyngeus sphincter (Fig. 341). The explanation of the underlying cause of the diverticulum put forward by Negus, seems to be the most likely one. Negus has drawn attention to the fact that in animals which climb trees, or in man who has a common origin with this type of animal, the crico-pharyngeus is very well developed. This is to prevent air entering the gullet when the glottis is closed preparatory to making the strong muscular effort associated with the climbing of a tree. In the view of Negus the underlying cause of the diverticulum is spasm of the crico-pharyngeus muscle. This impedes the entering of food into the upper œsophagus and so throws a strain on the posterior wall of the pharynx with the ultimate production of a diverticulum. The diverticulum starts as a bulge between the muscle fibres and with the entry of food and air the bulge becomes an actual sac, and as this sac enlarges it tends to move to the side of the neck, usually the left, and also to descend As the sac enlarges and descends the orifice of the diverticulum becomes rotated so that eventually it lies in a horizontal plane. When this stage has been reached the orifice of the diverticulum lies in the same line as the pharynx, and the œsophageal opening becomes displaced forwards so that food or instruments passed through the mouth tend to enter the diverticulum rather than the œsophagus.

Symptoms. At first the symptoms are subjective and the patient may feel that food sticks in the throat or sometimes a crumb will remain at the back of the throat until it is dislodged by hawking and coughing. At this stage the symptoms are principally due to the associated crico-pharyngeal spasm Later when the sac attains a greater size, it may fill up with food during the course of a meal, and dysphagia may occur towards the end of the meal owing to the pressure of the contents of the diverticulum on the œsophagus. Fullness of the neck may be noticed and pressure on the neck may cause the sac to be emptied so that food comes back into the mouth As the condition advances and rotation of the orifice occurs the neuro-muscular mechanism is interfered with and the dysphagic symptoms become more marked With a pouch of this size the mixture of food and air may give rise to gurgling sounds which are particularly noticeable on drinking and are often heard by the patient's friends. From time to time the sac may become inflamed and give rise to pain in addition to the symptoms already described. A carcinoma sometimes develops in a longstanding diverticulum.

Treatment. The first line of treatment is to carry out thorough dilatation by means of a Plummer or Negus hydrostatic bag (Fig. 340). This can be done conveniently at the time of the œsophagoscopic examination. To make sure that the dilatation is effective

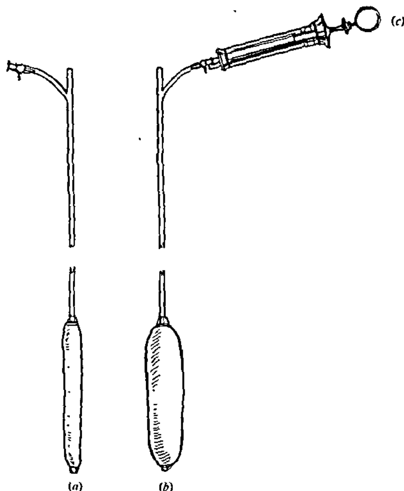


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Treatment. When the sac is of small size and most of the symptoms seem to be due to the associated spasm, it may be sufficient to dilate the crico-pharyngeus first at œsophagoscopy and later by instructing the patient to swallow a Hurst mercury bougie.



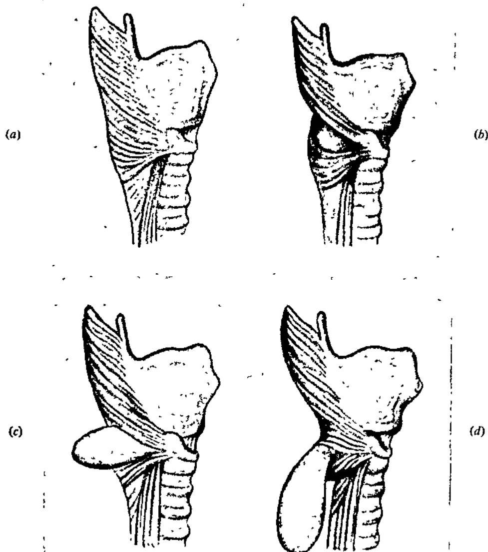
(*From the "Surgery of the Œsophagus," by R H Franklin, Edward Arnold & Co*)

FIG 342 Pharyngeal diverticulum in a man of seventy-five.

Any iron deficiency should be corrected

In more advanced cases the sac itself should be removed. The patient is admitted to hospital a few days before operation so that the sac can be washed out and antibiotics administered.

Diagnosis. The history is usually very suggestive and the diagnosis is confirmed by X-ray examination. A high stricture may cause confusion but in the case of a pouch the lower part fills up first and the opaque material spills over from the top, and this should serve to distinguish the two conditions (Fig. 342).



(From the "Surgery of the Œsophagus," by R. H. Franklin, Edward Arnold & Co.)

FIG. 341. Pharyngeal diverticula (after Negus)
(a)
(b)
(c)
(d)

Direct inspection with the œsophagoscope shows the size of the sac and excludes the presence of inflammatory or neoplastic changes. Instrumentation is potentially very dangerous because, as has been pointed out above, the instrument tends to pass more readily into the diverticulum than into the œsophagus and if this danger is not recognized perforation may occur.

produced by some adjacent inflammatory lesion, and once herniation has occurred food tends to enter the sac and cause the pouch to enlarge. Occasionally there may be associated cardiospasm.

Symptoms. These are often absent and the presence of a diverticulum may be discovered during the course of an X-ray examination carried out for other reasons. Sometimes dysphagia and pain are complained of.

Treatment. If no symptoms are present, treatment is not required. When there is associated narrowing of the œsophagus at the level of the diverticulum, dilatation should be carried out. If the presence of food in the pouch or attacks of inflammation give rise to symptoms, operative treatment may be required. When operation has to be carried out, repair is effected through a thoracotomy incision. For pouches in the lower third of the œsophagus it is better to approach through the left side of the chest, whereas those situated in the middle third are readily exposed by a right thoracotomy incision. At operation it may be sufficient to invert the mucous membrane which is protruding and repair the defect in the muscle coats with interrupted sutures of fine silk. If the sac is large, it may require excision and meticulous care must be taken in repairing the œsophagus in order to minimize the risk of any leak. The pleural cavity should be drained into a water-seal bottle for 48 hours after operation.

SIMPLE STRICTURES OF THE ŒSOPHAGUS

Congenital strictures of the œsophagus are extremely rare and many of the so-called congenital strictures are really the result of reflux œsophagitis. Similarly in adult life acquired simple strictures may be the result of this condition, the treatment has been considered above. Simple strictures resulting from corrosives can normally be treated by dilatation but this should not be started until 8 weeks have elapsed since the injury for fear of damaging the injured tissues. If the patient is seen immediately after a corrosive injury, it is important to administer suitable demulcents and antidotes, and if the injury is very severe to carry out a temporary gastrostomy for the purpose of feeding. After an interval of 8 weeks, or if the patient is seen at a later stage when a stricture has already developed, auto-dilatation using gum-elastic bougies is the method of choice. The nature and extent of the stricture must be accurately diagnosed in the first instance by radiological and œsophagoscopic examination, and any complicating feature such as the presence of a fistula, excluded. Auto-dilatation in which the patient is taught to swallow a gum-elastic bougie is a safe method provided no force is used. It has the great advantage over œsophagoscopic dilatation in that the procedure may be carried out at daily intervals and many patients in whom œsophagoscopic dilatation has failed can be restored to normal swallowing by this method.

SPONTANEOUS RUPTURE OF THE ŒSOPHAGUS

This term should be reserved for the rupture of a previously healthy gullet. A famous example was recorded in 1723 when Baron De Wassenaer, Grand Admiral of Holland, died of the condition. The subject was reviewed by Barrett in 1946 and he was the first surgeon to treat the condition successfully by operation. This case was reported in 1947.

The rupture normally always occurs at the lower end of the œsophagus. The

Early operations for this condition were fraught with danger because of the risk of mediastinal infection which sometimes led to the death of the patient. Leakage at the suture line if not followed by fatal infection, sometimes gave rise to a persistent and troublesome fistula. To overcome these dangers and difficulties the two-stage operation was devised. At the first operation the fundus of the sac was drawn upwards above the opening into the pharynx and retained in this position by stitches. The idea was to keep the sac empty and thereby relieve the patient's symptoms. The second stage consisted in excising the sac should this prove necessary. The trouble occurred if the second stage was called for because it was then found that adhesions to the surrounding tissues made removal of the sac extremely difficult.

With proper attention to the preparation of the sac, meticulous surgical technique and an understanding of how to make the anastomosis the early pitfalls can be avoided, and if operation is called for the one-stage procedure should be used.

Operation. General anæsthesia is administered through an endotracheal tube and the patient's head turned well to the right and a small sandbag placed under the left shoulder. The incision which gives the best exposure is one passing along the anterior border of the sternomastoid and extending from the level of the angle of the jaw to the sternoclavicular joint. The incision is deepened and the sternomastoid freed and retracted outwards. If the exposure is not satisfactory, the anterior half of the sternomastoid is divided immediately above the sternal attachment. The thyroid gland is retracted towards the opposite side and the carotid sheath retracted outwards. The inferior thyroid vessels may have to be divided The retro-pharyngeal space is opened up by blunt dissection with the finger and the sac identified. To help in the identification of the sac it is a good plan at this stage to pass a large stomach tube through the mouth and down the pharynx. The tube will usually pass into the diverticulum and when this has been identified the tube is withdrawn a little way and then guided down the œsophagus. The tube serves a useful purpose at a later stage in the operation in determining the level at which to divide the sac. Unnecessary handling of the sac is avoided by the introduction of fine stay sutures. The muscular coat is usually absent over the fundus of the sac but is present around the neck. This muscle should be carefully preserved in the form of a cuff so that when the mucous membrane has been divided and sutured the muscle can be sewn over the suture line. The sac is divided by stages and the opening sutured carefully with fine catgut as the division proceeds. The muscle is used to cover the mucous membrane and kept in place with interrupted sutures of fine silk. To prevent recurrence it is a good plan to introduce two or three sutures to hold the pharynx back to the prevertebral fascia. A soft drain is placed in the retro-pharyngeal space and care taken that there is no pressure on the suture line itself

After-treatment. Antibiotics are administered and fluid requirements given intravenously Nothing by mouth should be allowed for 3 days excepting sips of boiled water. On the third day liquid food is given and this is gradually increased until soft solids are taken after a week and a normal diet resumed about a fortnight after operation. The drain is removed on the fourth day.

DIVERTICULA OF THE THORACIC ŒSOPHAGUS

These occur most often in the distal quarter of the thoracic œsophagus. The condition is very rare. The cause of a diverticulum in this situation is often the traction

due to reflux œsophagitis in the presence of varicosities, and the operation of partial gastrectomy often improves the œsophagitis and in this way may lessen the tendency to bleed.

Treatment Aimed at Diminishing the Risk of Further Hæmorrhage

In patients who have survived a massive hæmorrhage it may be necessary to consider treatment aimed at preventing a recurrence. The appropriate operation is often difficult to decide because so many factors must be considered and an attempt must be made to differentiate patients in whom the obstruction is intra-hepatic from those in whom there is an extra-hepatic cause. The latter group is more likely to benefit by surgery.

NEW GROWTHS OF THE ŒSOPHAGUS

Simple Tumours

Simple tumours of the œsophagus are very rare and are unimportant in comparison with carcinoma. All types of simple tumour occur and they may arise from any of the coats of the œsophagus. Myoma, leiomyoma, lipoma, fibroma and hæmangioma have all been described and in addition to these, aberrant thyroid tissue may be found in the œsophagus. Epithelial and dermoid cysts also occur. Some of these tumours have been found at autopsy, never having given rise to symptoms during life. In other cases dysphagia leads to a diagnosis of carcinoma and when œsophageal cysts are present enlargement outwards into the pleural cavity may produce the signs of an empyema. Occasionally a simple polypoid growth in the œsophagus develops a long stalk which may cause remarkable symptoms such as extrusion of the polyp from the mouth, or death by suffocation from blockage of the entrance of the larynx.

Malignant Tumours

Sarcoma and melanotic sarcoma have been described but both conditions are of extreme rarity. Carcinoma of the œsophagus, on the other hand, is relatively common and if the cardiac end of the stomach is included the total number of cases comprises a very considerable proportion of the total number of cancers of the alimentary tract. Some 2,000 people die annually in Great Britain from the condition. The commonest form is a squamous-celled growth, but occasionally basal-celled carcinoma occurs. When the growth has started in the stomach the histology shows adenocarcinoma. Twenty-five per cent of cancers arise in the upper third of the œsophagus and hypopharynx, 30 per cent in the lower third and 45 per cent in the middle third. The condition is more common in men than in women, 80 per cent occurring in the former. Post-cricoid cancer is, however, more usual in women and it may occur as a sequel to the Plummer-Vinson syndrome. No age is exempt but most of the patients are between 50 and 70 years of age. In approximately a third of the patients early dissemination occurs to the hilar lymph nodes, [illegible] from the [illegible] involve vital structures such as the lung, bronchus, or great vessels so that pulmonary complications or a major hæmorrhage may cause sudden death. In the absence of such a termination the progress of the malignant stricture first prevents the swallowing of solid food, then liquids, and finally the patient is unable to swallow saliva.

Diagnosis. Dysphagia is the common presenting symptom although this may be

occurrence of the rupture is usually, but not always, associated with alcoholic excess accompanied by violent vomiting and the immediate cause of the rupture is probably the result of inco-ordinated muscular action of the œsophagus in the presence of excessive intraœsophageal pressure associated with the violent vomiting. Crush injuries sometimes produce this type of injury.

Clinical Features. The onset is sudden and is characterized by violent pain which simulates a ruptured peptic ulcer of the duodenum or stomach. The patient is shocked and usually cyanosed. A distinguishing feature is the excessive thirst of which the patient complains. This, together with the cyanosis and grunting respiration, should serve to raise the possibility of a ruptured œsophagus. Later surgical emphysema appears at the root of the neck but an attempt should be made to diagnose the condition before this occurs. X-ray examination of the chest may disclose emphysema in the posterior mediastinum and fluid or gas in the pleural cavity and so make an early diagnosis possible. Aspiration of fluid from the pleura confirms the diagnosis if blood and gastric contents are recovered.

Treatment. The condition is urgent and thoracotomy and closure of the perforation must be carried out and water-seal drainage of the pleural cavity instituted. Antibiotics are administered.

ŒSOPHAGEAL VARICES

Œsophageal varices are one of the most important complications of portal hypertension and bleeding from these varices accounts for over half the deaths in patients suffering from this condition.

Signs and Symptoms. The hæmorrhage may be slight and may occur over a long period, or may be copious and catastrophic and simulate the bleeding which occurs from a peptic ulcer of the stomach or duodenum. In the case of bleeding from œsophageal varices the patient may complain of a feeling of heaviness behind the sternum which is followed by an effortless regurgitation of bright red blood. Shortly afterwards nausea may be complained of and the patient then brings up altered blood mixed with gastric contents and clots. If a history such as this can be obtained, it serves to distinguish the bleeding from that due to a peptic ulcer.

Treatment for Massive Hæmorrhage. Blood transfusion may be essential but over-transfusion increases the tendency to further hæmorrhage. Pressure may be applied to the bleeding varices by introducing a Miller-Abbott tube so that the bag lies at the level of the cardia The bag is distended and the patient is fed through the lumen of the tube.

Sometimes it is impossible to differentiate between bleeding from varices and bleeding from a peptic ulcer and an urgent operation is undertaken. In these circumstances a gastrectomy may be carried out even though no ulcer can be felt, because it is a matter of common experience that a small peptic ulcer may be impossible to feel from the outside of the stomach and a search through a gastrostomy wound may be obscured by the contained blood. When a partial gastrectomy has been done under these circumstances and no peptic ulcer is subsequently discovered, the patient nevertheless stops bleeding. It may be that this treatment has proved effective because the left gastric vessels are ligatured during the course of the operation and the removal of part of the stomach may have lessened the chance of subsequent hæmorrhage The long-term improvement following this successful intervention may be due to the fact that the bleeding in these patients is partly

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problem in the days when anæsthesia had not reached the high standard expected today and when antibiotics were not available.

At this time but not generally known in the Western World, Ohsawa in Japan was



(From the "Surgery of the Œsophagus" by R. H. Franklin. Edward Arnold & Co.)

Fig. 343 Extensive carcinoma of the lower œsophagus

carrying out a type of operation very similar to that used for the lower end of the œsophagus today. Writing in 1933 he described his experiences with 101 cases over the preceding 7 years. These patients were suffering from carcinoma involving the cardia and, using a left-sided abdomino-thoracic approach, he was able to resect 18 with

preceded by fatigue and vague ill-health or "indigestion," and occasionally hæmorrhage may draw attention to the condition. The dysphagia is usually progressive but sometimes a temporary remission causes difficulty in diagnosis. The length of history may also cause difficulty because cancer may arise in a patient who has been suffering from cardiospasm for many years.

General Examination. This should be carried out with care but nothing abnormal may be found apart from the loss in weight. Sometimes enlarged cervical lymph nodes are felt or abdominal examination shows an enlarged liver or a growth arising in the stomach. Rectal examination may reveal secondary nodules from a carcinoma of the cardiac end of the stomach. The vocal cords should be inspected for paralysis due to direct involvement of the recurrent laryngeal nerve or to involvement by a malignant lymph node. The patient should be examined eating and drinking to assess the dysphagia, and if a cough is produced on drinking the reason may be that the air passages are invaded and further investigations should be made to confirm this suspicion.

Radiological Examination. This must always be carried out before any instrument is passed. Sometimes the cause of the dysphagia proves to be extrinsic, for example, by pressure from an aneurysm or from a new growth of the lung. X-ray examination is a very reliable method of diagnosis in the middle part of the œsophagus, but, at the upper end there is a tendency to diagnose a growth in the hypopharynx when, in fact, the patient is suffering from crico-pharyngeal spasm. At the lower end of the œsophagus an extensive carcinoma of the fundus of the stomach may be masked by an overlying spasm, so that the patient is wrongly diagnosed as suffering from cardiospasm. In a typical case a stricture produced by malignant disease is irregular and in many cases the lumen is eccentric (Fig. 344) and contrasts with the smooth appearance of cardiospasm. Whereas the hold-up in the case of cardiospasm is always at the level of the diaphragm, malignant disease may occur in any part of the œsophagus. Dilatation of the gullet above the growth is not common, but if a carcinoma occurs in a patient who has been suffering from cardiospasm dilatation may be marked and give rise to errors in diagnosis. Reflux œsophagitis sometimes produces gross distortion of the lower end of the œsophagus.

The X-ray findings should be confirmed by direct examination with the œsophagoscope and a biopsy made unless there is some absolute contra-indication to this procedure. Œsophagoscopy is particularly important if there is no X-ray explanation for the patient's dysphagia.

Treatment of Carcinoma of the Œsophagus

Resection is still the best method of treatment if practicable, and death from metastases is less unpleasant than death from the original growth.

Historical Note. The cervical œsophagus was successfully resected by Czerny in 1877. The cardia was resected by the abdominal approach by Voelcker (1908), Kummel (1910) and Bircher (1918) The thoracic œsophagus was not attacked successfully until 1913 when Torek operated on a growth in this situation and the patient lived for 13 years without recurrence No further success was reported until Grey Turner was able to describe a case in 1933 in which the "collo-abdominal" or "pull-through" method was used. This method consisted in mobilizing the thoracic œsophagus by means of a finger introduced through the œsophageal hiatus from below and another finger through the thoracic inlet from above This was an ingenious method of dealing with a difficult

problem in the days when anæsthesia had not reached the high standard expected today and when antibiotics were not available.

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immediate anastomosis, and 8 of these patients survived. Ohsawa was displaying great vision because the few contemporary surgeons who were attacking the condition aimed at first removing the œsophagus, leaving the patient with a cervical œsophagostomy and a gastrostomy, with the hope of constructing an ante-thoracic œsophagus should the patient survive.

The first successful removal, with immediate reconstruction, in the Western hemisphere was described by Adams and Phemister in 1938. The surgical management of cancer of the lower end of the œsophagus now entered a more promising period, at any rate from the point of view of immediate mortality, and Garlock, Churchill and Sweet, showed that successes could be obtained in a reasonable proportion of cases. Growths of the middle third proved more difficult and Ivor Lewis in 1946 described a series of patients in which an abdominal mobilization of the stomach was followed by a right trans-pleural approach to the growth.

Opinions vary at the present time as to the type of operation suitable in particular instances and the operations preferred by the author are discussed below.

The author's figures of the immediate mortality and operability rate over a 5-year period are shown in the following table:

CANCER OF THE THORACIC ŒSOPHAGUS AND CARDIA, 1951–5

	Number of Cases	*Deaths*
Resection of growth and anastomosis of stomach to cervical œsophagus or pharynx	2	2
Anastomosis of stomach to cervical œsophagus to short-circuit the growth at the level of the thoracic inlet	1	—
Resection of growth and anastomosis in the thorax above arch of aorta	17	6
[illegible] arch of aorta	25	11
[illegible]	7	2
[illegible]	1	1
Palliative short-circuit of middle of œsophagus	12	3
Souttar tube	3	1
Gastrostomy only	3	2
Jejunostomy only	1	1
Exploration only	7	2
No operation	8	6
	87	37

Preliminary Preparation. Before undertaking any major operation on the œsophagus it is essential to prepare the patient thoroughly, and it is largely due to the recognition of this fact that the immediate mortality has been lowered in recent years. The first essential is to try to improve nutrition and this can be achieved by giving the patient a fortified liquid diet, the basis of which is milk and to which should be added eggs, sugar, butter and dried milk. This fluid is not very palatable and in addition fruit juices, alcohol and soup may be given. Vitamins are added to the diet or given by injection. All solids are removed from the dietary. It is essential to stress this point because after a few days the patient finds that swallowing is easier and there may be a natural desire to try some easy solid food, the only effect of which is to block up the narrowed lumen of the œsophagus. Another important point to impress on the patient is that all ideas of regular meal times should be abandoned and he should be encouraged to take the liquid at frequent intervals

during the day so that the total intake is adequate. Four or five pints in 24 hours should be the aim.

During this period of preparation the patient is encouraged to take exercise when he feels strong enough to do so and he should carry out regular breathing exercises under proper supervision. An assessment of the state of the heart and lungs is made and if necessary postural drainage instituted. Septic teeth are cleaned and an attempt made to improve the state of the gums by massage with salt and water. This is a better plan than embarking on large-scale dental extractions although loose teeth may need to be removed. The sipping of dilute hydrogen peroxide solution from time to time is beneficial in diminishing infection in the neighbourhood of the growth and in removing debris. It is important to make sure that the patient is not suffering from impacted fæces as the result of the preceding prolonged starvation. The co-operation of the patient can usually be gained in all these measures if the importance of them is pointed out.

OPERATIVE PROCEDURES FOR GROWTHS OF THE THORACIC ŒSOPHAGUS

(1) **For Growths Involving the Cardia** (Fig. 344). The stomach, spleen, tail of pancreas and the great omentum, together with the lower 2–3 in. of the œsophagus, are removed in one block as described by Allison. Continuity is restored by means of a Roux en Y anastomosis.

The operation is carried out through a left abdomino-thoracic approach, removing the eighth rib and extending the incision forwards to divide the costal margin. The diaphragm is incised in a radial manner from the periphery down to the œsophago-gastric junction, dividing the right crus. The spleen is held up into the wound and the lieno-renal ligament divided. The great omentum is drawn upwards and the bloodless fold connecting it with the transverse colon displayed and divided. The spleen and stomach are now drawn over towards the patient's right-hand side and the tail of the pancreas with the splenic vessels brought into view. These structures are divided and the splenic vessels ligatured. Any lymph nodes which can be seen lying along the upper border of the pancreas are dissected free. The left gastric vessels are brought into view and are ligatured and divided. The right gastro-epiploic vessels and the right gastric vessels are divided and clamps applied to the first part of the duodenum, which is divided and the cut end closed and invaginated. The stomach, together with the spleen and tail of the pancreas, is turned up into the chest and the attachments holding the cardia to the posterior part of the œsophageal hiatus are divided and the mobilization of the œsophagus is continued for 2 or 3 in., taking care to divide both vagi. Stay sutures are introduced at the proposed line of section of the œsophagus which is then divided in the manner described on page 661. Continuity is restored by means of a Roux en Y anastomosis and the operation is completed as described on pages 660 and 661.

(2) **For Growths not Involving the Cardia but which lie below the Level of the Arch of the Aorta** (Fig. 346). Growths in this situation are conveniently dealt with by resecting the œsophagus from a point above the arch of the aorta to the cardia and using the stomach to restore continuity. The operation is conveniently carried out from the left side, excising the eighth rib and gaining further room by drawing the scapula forwards and making another incision in the fourth left intercostal space. The operative technique is

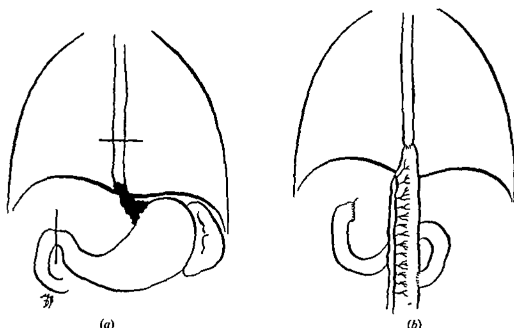


FIG. 344 (a) Carcinoma involving the cardiac end of the stomach and the lower end of the œsophagus. The resection includes the lower part of the œsophagus, the whole of the stomach, spleen, the tail of the pancreas and the great omentum (b) Continuity has been restored by a Roux en Y anastomosis

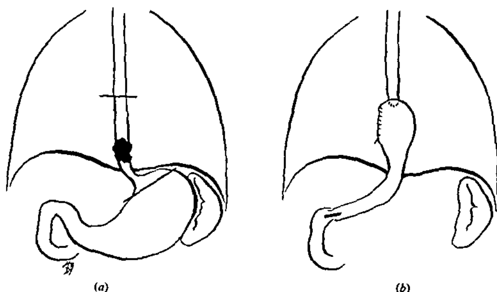


FIG 345. (a) The growth is situated in the œsophagus below the arch of the aorta and does not involve the cardiac end of the stomach The stomach is mobilized as described in the text and the œsophagus resected from above the aortic arch The cardiac end of the stomach is included in the resection (b) The mobilized stomach is anastomosed to the œsophagus above the arch of the aorta The pyloric sphincter is divided

precisely the same as has been described for resection of the œsophagus in extensive œsophagitis (page 662).

(3) For Growths Involving the Œsophagus at the Level of the Arch of the Aorta (Fig. 346(*a*), (*b*)). In this situation the growth will almost certainly involve the right pleura and possibly the vena azygos arch as well. For this reason mobilization from the left side of the chest is certain to open both pleural cavities and may, in addition, be followed by dangerous hæmorrhage from the vena azygos arch. It is therefore better to resect the œsophagus in these cases from the right side of the chest, having previously mobilized the stomach through a midline abdominal incision.

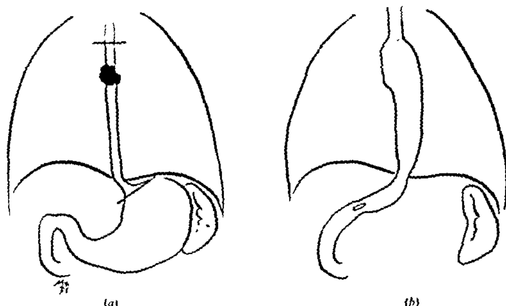


Fig. 346 (*a*) The growth is situated at the level of the aortic arch and is going to be freed by a right-sided thoracic approach after preliminary mobilization of the stomach by the abdominal route (*b*) Through a right-sided thoracic approach the mobilized stomach has been drawn high up into the thorax to be anastomosed to the stump of the œsophagus The pyloric sphincter has been divided

With the patient lying on his back, a midline abdominal incision is made extending from the xiphisternum to the umbilicus, and the stomach is mobilized by dividing the vasa brevia and the gastrocolic omentum halfway to the pylorus, care being taken to preserve the vascular arch on the side of the stomach. Should any difficulty be encountered with the spleen owing to adhesions or a tear producing hæmorrhage, it is better to remove the spleen at this stage. The left gastric vessels are ligatured and divided and the mobilization of the stomach is completed right up to the diaphragm, and to achieve this it may be necessary to divide the left coronary ligament of the liver. The pyloric sphincter is divided without opening the mucous membrane and the abdomen closed. The patient is turned on to his left side and the thorax opened by resecting the fifth rib. The vena azygos arch is ligatured and divided and it may be necessary to remove part of the arch with the underlying growth. The œsophagus is mobilized from a point about 1½ in from the thoracic inlet right down to the diaphragm The mobilized stomach is then drawn up into the chest. The œsophago-gastric junction is divided and the gastric side closed and inverted. An opening is made in the fundus of the stomach corresponding exactly in size with the cut end of the œsophagus. The stomach and œsophagus are

then anastomosed in exactly the same way as has already been described in the case of œsophago-gastrostomy for œsophagitis on pages 661 and 662. The fact that the procedure is being carried out from the right side makes no difference to the details of the anastomosis. A water-seal drain is introduced through a separate intercostal incision and the wound closed.

(4) **For Growths situated in the Thoracic Œsophagus above the Level of the Arch of the Aorta** (Fig. 347 (*a*), (*b*)). This is one of the most unfavourable situations in which a growth can occur because it is not possible to carry out an adequate resection and anastomosis without bringing the stomach through the chest right up into the neck, and few patients

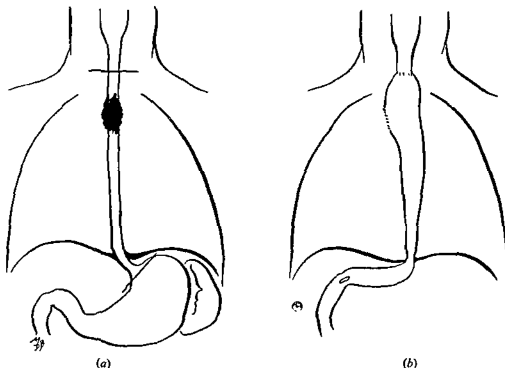


FIG. 347 (*a*) The growth is so high in the thorax that anastomosis can only be satisfactory if it is made in the cervical region. Through a left-sided thoraco-abdominal approach the stomach is mobilized, the pyloric sphincter divided and the œsophagus freed throughout its entire thoracic course (*b*) A cervical incision is made and the fundus of the mobilized stomach anastomosed to the stump of the cervical œsophagus or pharynx

are fit enough to withstand this procedure. It is in this situation that there may be a useful field for the employment of a polythene tube to bridge the gap of a limited resection, but the author's experience with this type of operation has not been encouraging as a leak is very liable to occur. In those patients who are not fit to stand the major procedure of resection with cervical œsophago-gastrostomy, it is often wise to be content with introducing a Souttar's tube. The introduction of a Souttar's tube is described under palliative procedures.

If the patient is fit for the major procedure, the stomach is first mobilized from the abdomen as has been described on page 662. The patient is now turned on to his right side, the seventh rib resected and the whole of the thoracic œsophagus mobilized. The stomach is drawn up into the chest and a strong stitch introduced through the fundus. The ends of this stitch are brought out just below the inner end of the left clavicle so that the stomach can subsequently be recovered in the neck. The chest wound is closed in the

usual way after the introduction of a water-seal drain. The patient is now turned on to his back again and an incision made along the anterior border of the left sternocleidomastoid. The muscle is divided just above the clavicle to give good access to the cervical œsophagus, which is exposed by dissecting in the space between the thyroid gland on the inner side and the great vessels on the outer side. It may be necessary to divide the inferior thyroid artery. The cervical œsophagus is mobilized with the finger and the fundus of the stomach is hooked up in the neck. The requisite amount of cervical œsophagus is excised and the fundus of the stomach anastomosed either to the upper part of the cervical œsophagus or to the pharynx itself. The author has been able to carry out this anastomosis successfully on three occasions. A soft latex drain is placed near the anastomosis and the incision in the neck closed.

Technical Considerations in Carrying out the Above Operations

Anæsthesia should be administered through an endotracheal tube so that during the operation the lungs may be fully expanded from time to time. It is particularly important to see that full expansion occurs before the chest is closed and in all cases a water-seal drainage tube must be introduced. Clamps must never be applied to any part of the œsophagus which is going to be used in the anastomosis. The blood supply of the œsophagus is satisfactory provided that undue mobilization is not carried out, and care should be taken that only sufficient œsophagus is mobilized above the line of section to effect a satisfactory anastomosis. It must be remembered that there is practically no extensibility of the œsophagus, and for this reason the stomach or small bowel which is being used to restore continuity must always be brought up to the œsophagus.

Sutures are very liable to cut out in the œsophagus and the author has found that a very satisfactory method of overcoming this is to introduce three or four interrupted mattress sutures of linen thread in the posterior part of the anastomosis. These sutures pass through all coats of both œsophagus and stomach, or bowel as the case may be. A continuous catgut suture is now introduced through all coats and when the suture reaches the anterior part of the anastomosis it is a help if the needle is passed from the mucous surface on each side, as by this means it is made certain that all coats are included. It is impossible to introduce a satisfactory Lembert layer and attempts to do so often produce damage to the œsophageal muscle It is better to concentrate on one or two "special" stitches and to reinforce the suture line with omentum or pleura. At one end of the suture line a "special" stitch is introduced first into the stomach and then into the pleura over the aorta at a slightly higher level than the anastomosis. From there the stitch passes into the muscular wall of the œsophagus just above the anastomosis and then back into the stomach When this stitch is tied it draws a peritoneal covered surface of stomach or bowel over the end of the anastomosis and at the same time takes the strain off the suture line. A similar stitch is introduced at the other end of the suture line but here the base of the pulmonary ligament has to be used instead of the pleura over the aorta. This stitch has to be modified according to the level at which the anastomosis is being made, but the principle of relieving the suture line of any strain can be adhered to. The rest of the suture line can be covered with a convenient fringe of omentum or a flap of pleura may be turned over and stitched in position. It is important to attach the stomach or bowel to the parietal pleura at several points. The opening in the diaphragm requires particular attention and the cut edge of the diaphragm must be very carefully stitched to the stomach

or bowel in order to prevent the risk of any herniation. The remainder of the incision in the diaphragm should be sutured with interrupted thread and in addition a continuous catgut suture to prevent any risk of bleeding.

Palliative Procedures

A major exploration is unjustifiable if there is clear evidence that the growth cannot be resected completely. Such evidence may be present in the form of paralysis of the vocal cord even though the primary growth is situated low in the œsophagus, or there may be evidence of involvement of the trachea or bronchi, or obvious metastases in the liver

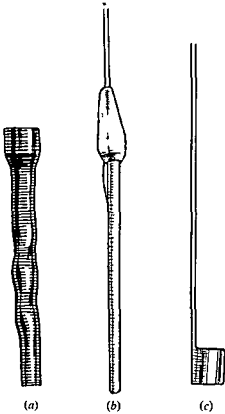


Fig. 348 (*a*) Souttar's tube of German silver wire (*b*) Introducer to carry the tube into its proper position (*c*) Ring used to hold the tube in position while the introducer is withdrawn

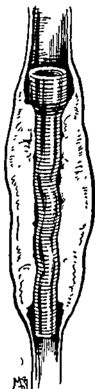


Fig 349. Souttar's tube correctly in position, with the shoulder of the tube resting on the upper part of the growth.

or in the lymph nodes at the root of the neck. In these circumstances or in cases where the cardiac or pulmonary condition of the patient is such that a major operation is not *justifiable, a palliative procedure should be carried out.*

Intubation by Means of a Souttar's Tube. This will often provide considerable relief and even though the tube may tend to slip through the malignant stricture and necessitate replacement by a larger one, the patient may continue for several months in comparative comfort, taking an almost normal diet.

The patient is anæsthetized and the growth inspected through a large bore œsophagoscope. The lumen is identified by means of a gum-elastic dilator and dilated cautiously so that it will receive the Souttar's tube. The tube itself is introduced with a guide which

passes beyond the growth and is held in position by means of a special introducer while the guide is withdrawn. As large a Souttar's tube as can be introduced should be used (Fig. 348).

The tube itself consists of a spiral of German silver wire, the upper end of the tube being expanded into the form of a collar which prevents it from slipping through the malignant stricture, and the fact that the tube is flexible enables it to adapt itself to any irregularities produced by the growth (Fig. 349)

It must be realized that the introduction of a Souttar's tube is accompanied by a *considerable risk of producing hæmorrhage or perforation and the method should only* be used in inoperable cases.

When the tube has been introduced, the patient should be instructed to mince his food carefully and to follow every meal with a drink of water. He should also sip dilute hydrogen peroxide solution several times a day to keep the tube clear.

Irradiation. Deep X-ray therapy applied in the form of rotation therapy sometimes produces considerable temporary relief, but there is a risk that the formation of a fistula into the œsophagus or trachea may be hastened by this treatment and for this reason it should not be advocated excepting as a last resort.

Limitation of the diet to a fortified liquid as has been described in the section on the preparation of the patient for operation, is in itself a valuable palliative procedure and may postpone the time when dysphagia becomes complete.

Gastrostomy is not a satisfactory palliation but it must sometimes be resorted to when the patient is unable to take fluids The rest afforded to the œsophagus by the gastrostomy will often enable mouth feeding with liquids to be resumed after a few days.

Palliative Procedures which may be used when Inoperability has been Determined at Exploration

If a major exploration has been undertaken and the condition is found to be too advanced for adequate removal, or if involvement of the root of the lung or aorta makes resection too hazardous, the best procedure to carry out is that of making a short circuit. The necessity for a short circuit most often arises when the growth is at the level of the root of the lung. The growth may be quite small but local involvement may make excision dangerous. *Under these circumstances a short circuit can be conveniently* carried out by mobilizing the stomach and freeing the œsophagus as far as the lower limit of the growth The fundus of the stomach is then brought to the œsophagus above the arch of the aorta. The mobility of the freed stomach enables this to be carried out without difficulty. A lateral anastomosis is made between the fundus of the stomach and the œsophagus.

A short-circuit may be impossible to carry out if the growth is situated high in the thorax. In these circumstances it may be possible to introduce a Souttar's tube, whereas it would have been impossible without the guidance afforded by a direct exposure

POST-OPERATIVE MANAGEMENT OF ŒSOPHAGEAL OPERATIONS

Care of the Chest

This starts at the end of the operation when the anæsthetist aspirates the bronchial tree. Post-operative sedation aims at enabling the patient to breathe without undue pain

but avoiding large doses which suppress respiratory movements. With this in view small repeated injections of pethidine or morphia are used.

An oxygen tent is often necessary for a few hours.

Breathing exercises are started as soon as the patient is fit enough and the ward nurse must always stress the importance of proper breathing and the expectoration of any accumulated secretions and not leave these instructions only to the physiotherapist on the occasion of her visit to the ward. Holding the patient's chest firmly while he breathes and coughs often gives him confidence and lessens the pain. Occasionally bronchoscopic aspiration is required.

The antibiotics which were started before the operation are continued for a week post-operatively.

X-ray Control of the Chest

For the first few days X-ray examinations are made in case there should be an accumulation of air or fluid which has not been diagnosed clinically.

The water-seal drainage is checked frequently to make sure that it remains air-tight and that the column of fluid moves with respiration and maintains a negative pressure of 3–5 in. of water. Excessive movement of the column should be controlled by partial clamping, and if movements stop altogether the tube is "milked" to clear any blockage in the lumen. Sudden dyspnœa in the absence of any radiological explanation should lead to exploration with an aspirating needle. The intercostal drainage tube is usually left in position for 3 or 4 days.

Intravenous Therapy and Feeding

For the first 48–72 hours fluid requirements are maintained by intravenous therapy. A Ryle's tube is left in position at the operation with its tip beyond the anastomosis, and for the first 24 hours hourly aspiration is carried out to make sure that distension of the stomach or small bowel distal to the suture line is not taking place.

On the second day the tube may be used for feeding purposes and at the same time sips of half-strength saline are allowed by mouth. As a rule the Ryle's tube is removed within the first 48 hours. On the following day an ounce of milk is substituted for the saline and after this the diet is gradually increased so that the patient is taking a normal diet 10–12 days after the operation.

Complications

Leakage at the Anastomosis. If a leak in the anastomosis occurs, the chest drain must be kept in position or reintroduced and an intravenous drip set up once more to provide the patient's fluid requirements. If the leak occurs after the first week, it may be an advantage to continue feeding semi-solids by mouth, but should it occur in the first few days aspiration through the Ryle's tube is carried out at frequent intervals. A jejunostomy is required if the fistula shows no signs of closing after 2 or 3 days.

Hæmorrhage. This may occur from a bleeding point on the diaphragm and blood may pass either into the thorax or abdomen. If it passes into the latter, the diagnosis is often extremely difficult and this fact serves to emphasize the importance of making absolutely certain of complete hæmostasis during operation.

Small Bowel Strangulation. This may follow herniation either through the diaphragmatic incision or through the abdomen in the transverse mesocolon. Like the above,

this may be an extremely difficult complication to diagnose and may well prove fatal before the diagnosis is confirmed. For this reason efforts must be made at the time of operation to attach the bowel or stomach carefully at the points where it passes through the diaphragm and mesocolon and to leave no gap.

Reflux Œsophagitis. This is very liable to occur if the stomach has been anastomosed to the lower end of the œsophagus and for this reason this type of anastomosis should not be used.

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W[illegible] ique in Congenital Œsophageal Atresia with Œsophago-tracheal Fistula. *Anaesthesia*, 6, 30.

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